Neurology

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Anesthesia / Pharmacotherapy

Anesthesia

17125

- Local anesthetic systemic toxicity, a potential complication of epidural analgesia, can cause CNS overactivity (eg, perioral numbness, metallic taste, tinnitus) and generalized tonic-clonic seizures.

Delirium

21340

- Inadequate emergence from general anesthesia (ie, delayed emergence, emergence delirium) is typically due to residual effects of anesthetic/adjuvant medication.
- It is usually temporary and resolves with reassurance and reorientation, although urgent evaluation to exclude more serious conditions should be considered.

Dystonia

17119

- Dystonias are involuntary muscle contractions that often involve the small muscles of the head and neck; oculogyric crisis results in a forced upward gaze deviation.
- Dystonias typically occur as extrapyramidal symptoms of a high-potency, first-generation antipsychotic (eg, haloperidol, fluphenazine).
- Management includes benztropine and diphenhydramine.

16229

- Blepharospasm is a form of focal dystonia characterized by recurrent forceful contraction of the eyelid muscles.
- Bright lights may trigger symptoms, whereas touching or brushing the skin around the eye may terminate the spasm (ie, "sensory trick").

Metoclopramide

106811

- Drug-induced parkinsonism is most commonly caused by a dopamine receptor antagonist.
- Use of such medications is considered an absolute exclusion criterion for the diagnosis of idiopathic Parkinson disease.
- Discontinuation of the offending medication typically leads to the resolution of symptoms in a few weeks to months.

Tremor

19545

- Essential tremor most often presents as an action tremor of the hands that is not associated with other neurologic symptoms.
- First-line treatment includes propranolol, which exerts its effects due to peripheral beta-adrenergic receptor blockade, and primidone, a centrally acting anticonvulsant.

19547

- An enhanced physiologic tremor is typically a fine, fast, symmetric action tremor of the hands that increases with sympathetic activity.
- Certain medications (eg, selective serotonin reuptake inhibitors, beta agonists) can cause enhanced physiologic tremor, which generally improves with dose reduction or medication cessation.

Trigeminal neuralgia

3461

- Trigeminal neuralgia is characterized by short, episodic, severe pain usually along the V2 (maxillary) and V3 (mandibular) branches of the trigeminal nerve (CN V), often triggered by minor stimuli.
- Oxcarbazepine or carbamazepine is first-line therapy.

Fibromyalgia

3205

- Fibromyalgia is a pain syndrome that occurs most commonly in young to middle-aged women and is characterized by fatigue, widespread pain, and cognitive/mood disturbances.
- Amitriptyline is an effective initial therapy.
- Pregabalin, duloxetine, and milnacipran are alternate therapies for patients not responding to tricyclic antidepressants.

Congenital and developmental anomalies

Cerebral palsy

2443

- Cerebral palsy is a nonprogressive motor dysfunction; prematurity is the leading risk factor.
- The most common findings are gross motor delay in infancy and spasticity (eg, hypertonia, hyperreflexia).

21410

- Cerebral palsy is a nonprogressive neurologic injury that most commonly occurs in premature infants with periventricular leukomalacia.
- Findings can include motor delay, early hand preference, spasticity, and hyperreflexia.

Chiari malformation

16250

- Chiari I malformation is characterized by inferior displacement of the cerebellar tonsils through the foramen magnum, and syringomyelia is a commonly associated condition.
- Although frequently asymptomatic, presentation may occur in adolescence/adulthood with occipital headache exacerbated by activity and Valsalva maneuvers.

Developmental milestones

7741

- Normal development of an 18-month-old toddler includes speaking ≥3 words, walking independently, scribbling, and following a simple command.
- The inability to walk without support by this age is a sign of gross motor delay.

4874

- Two-year-old children with appropriate language development should combine words into 2-word phrases. In addition, a stranger should be able to understand at least half of a child's speech at age 2.

Fetal alcohol syndrome

2444

- Fetal alcohol syndrome due to in utero alcohol exposure classically presents with the triad of small palpebral fissures, smooth philtrum, and thin vermilion border.
- Patients also have poor growth and associated neurodevelopmental issues (eg, development delay, intellectual disability, hyperactivity, poor social skills).

Fragile x syndrome

2441

- Physical features of fragile X syndrome can include a long face, prominent chin and forehead, protruding ears, joint hypermobility, and macrocephaly.
- Associated developmental delays and attention deficit hyperactivity disorder are common, but life expectancy is normal.

2361

- Fragile X syndrome is characterized by neuropsychiatric symptoms (eg, developmental delay, autism) and dysmorphic features (eg, elongated face, deep-set eyes, large ears).
- Diagnosis is with FMR1 DNA analysis.

Friedreich ataxia

15219

- Friedreich ataxia is an autosomal recessive, neurodegenerative disease that typically presents in adolescence with progressive ataxia, dysarthria, scoliosis, and cardiomyopathy.
- Loss of vibratory sense and proprioception also occur due to degeneration of the dorsal spinal columns.

2439

- Friedreich ataxia is an autosomal recessive, neurodegenerative disease that typically presents in adolescence with progressive ataxia and loss of position and vibratory senses.
- Genetic testing reveals an excessive number of trinucleotide repeat sequences.

Galactosemia

3193

- Galactosemia is a metabolic disorder in which deficient galactose-1-phosphate uridylyltransferase prevents the reduction of galactose to glucose.
- The disorder typically presents with jaundice, vomiting, hepatomegaly, and possibly seizures (due to hypoglycemia) in the first few days of life.
- The diagnosis is supported by the presence of nonglucose reducing substances in urine, which suggests galactosuria.

Homocysteine

3687

- Homocystinuria is an autosomal recessive disorder caused by cystathionine synthase deficiency.
- In addition to Marfanoid body habitus, patients with homocystinuria also have intellectual disability, downward lens dislocation, and hypercoagulability.
- Treatment involves vitamin supplementation and antiplatelet or anticoagulation to prevent thromboembolic events.

Huntington disease

3954

- Huntington disease is a highly heritable progressive illness that presents with a combination of psychiatric (eg, depression), cognitive (eg, executive impairment, memory loss), and neurologic (eg, chorea) symptoms.

Intraventricular hemorrhage

3666

- Intraventricular hemorrhage is a common complication of prematurity and can present with acute neurologic changes (eg, seizure, apnea), a bulging fontanelle, and a rapidly increasing head circumference.

3657

- The primary risk factor for intraventricular hemorrhage is prematurity due to fragile germinal matrix vessels.
- Symptomatic newborns may have apnea, seizures, and a bulging fontanelle as well as signs of acute blood loss (eg, anemia, tachycardia).
- Cranial ultrasound is diagnostic.

18572

- Preterm newborns are at increased risk of developing intraventricular hemorrhage (IVH) due to the presence of the germinal matrix, a fragile, highly vascularized area in the brain.
- Because IVH can be asymptomatic, all preterm neonates born at <32 weeks gestation require screening head ultrasound.

Lesch Nyhan syndrome

3817

- Lesch-Nyhan syndrome is caused by a deficiency in hypoxanthine-guanine phosphoribosyltransferase.
- Neurologic findings begin with developmental delay and hypotonia in infancy and progress to chorea, dystonia, and spasticity in early childhood.
- Cardinal findings include self-mutilation and gouty arthritis.

Myotonic Dystrophy

3661

- Myotonic dystrophy is an autosomal dominant trinucleotide repeat disorder that most commonly presents in adolescence or early adulthood with progressive weakness (eg, facial and hand muscles), muscle atrophy, grip myotonia (delayed muscle relaxation), cardiac conduction anomalies, and gastrointestinal tract disturbances (eg, dysphagia).
- Testicular atrophy can also occur.

Malignant hyperthemia

15653

- Malignant hyperthermia is a genetic disorder associated with sudden-onset tachypnea, tachycardia, myoglobinuria, and masseter/generalized muscle rigidity following exposure to succinylcholine or a volatile anesthetic.
- Most cases arise during or shortly after induction, but symptoms are sometimes delayed until just after anesthesia cessation.

Microcephaly

18788

- Microcephaly (head circumference <3rd percentile) is usually benign in children with normal development and an otherwise normal examination.
- Benign familial microcephaly is a common cause, and measuring the parents' head circumferences supports the diagnosis when adult head size is similarly small.

18791

- Pathologic features of microcephaly (head circumference below the 3rd percentile) include neurologic abnormalities, dysmorphic features, and rapidly declining head circumference percentiles.
- The first step in evaluation is MRI of the brain to assess for structural, genetic/metabolic, or infectious conditions associated with inadequate brain growth.

Neimann Pick Disease

2445

- Niemann-Pick disease type A is due to sphingomyelinase deficiency and presents at age 2–6 months with areflexia, hepatosplenomegaly, a "cherry-red" macula, and developmental milestone regression.
- Although Tay-Sachs disease presents in a similar manner, hepatosplenomegaly and areflexia are not seen.

Neurofibromatosis

2669

- Neurofibromatosis type 1 (NF1) is an autosomal dominant neurocutaneous disorder associated with café-au-lait macules and axillary and inguinal freckling.
- Patients with NF1 have an increased risk of seizures, learning disabilities, and optic pathway gliomas.

16699

- Vestibular schwannomas present with hearing loss and imbalance.
- Bilateral, hereditary schwannomas are most often associated with neurofibromatosis type II.

4049

- Neurofibromatosis type 1 is an autosomal dominant disorder associated with café-au-lait macules, skinfold freckling, Lisch nodules, and peripheral nerve sheath tumors (neurofibromas).

3550

- Patients with neurofibromatosis type 1 are at increased risk for optic pathway gliomas, which can present with headache and decreased visual acuity.
- MRI of the brain and orbits evaluates for an intracranial mass.

Respiratory physiology

20276

- Apnea of prematurity is caused by immature respiratory centers in the pons and medulla and affects virtually all preterm infants born <28 weeks gestation.
- Diagnosis is clinical, and no additional testing is required in a well-appearing infant with a classic presentation in the first few days of life.

Rett syndrome

15794

- Rett syndrome is characterized by a period of normal development followed by regression of speech, loss of purposeful hand use, stereotypical movements, and gait disturbance.
- Other features include early deceleration in head growth (ie, microcephaly), breathing abnormalities, autistic behaviors, and seizures.

SIDS

3907

- Sudden infant death syndrome refers to the unexplained death of infants age <1.
- Modifiable risk factors include smoke exposure (prenatal and postnatal) and prone- and side-sleep positioning.

Scalp Hematomas

18604

- A cephalohematoma is a collection of blood between the skull and periosteum caused by the rupture of subperiosteal vessels during delivery.
- Examination reveals a firm, nonfluctuant scalp swelling that does not cross suture lines.
- Management is with reassurance and observation because most cephalohematomas resolve without intervention.

16407

- Subgaleal hemorrhage is caused by the shearing of veins between the dural sinuses and scalp due to scalp traction during delivery.
- Blood accumulates between the periosteum and galea aponeurotica, causing diffuse, fluctuant scalp swelling.
- Rapid hemorrhage expansion can lead to hypovolemic shock, disseminated intravascular coagulation, and death.

2472

- Cephalohematoma is a subperiosteal hemorrhage that presents in the first day of life with a firm, well-demarcated scalp swelling that does not cross suture lines.
- Forceps- or vacuum-assisted deliveries increase the risk of developing cephalohematomas.
- Management is with observation because most resorb spontaneously.

Sturge weber syndrome

18737

- Glaucoma in infants typically presents with tearing, photophobia, blepharospasm, and an enlarged cornea and globe.
- Patients with Sturge-Weber syndrome are at increased risk due to a congenital anterior chamber angle anomaly.
- Evaluation of glaucoma includes tonometry to measure intraocular pressure.

2763

- Sturge-Weber syndrome is a neurocutaneous disorder characterized by a capillary malformation (port-wine stain) along the trigeminal nerve distribution and leptomeningeal capillary-venous malformations that affect the brain and eye.
- Seizures, intellectual disability, and visual field defects can occur.

Tuberous sclerosis

18696

- Tuberous sclerosis complex (TSC) is an autosomal dominant neurocutaneous disorder characterized by distinct skin features and benign tumor formation, including subependymal giant cell tumors in the brain and cardiac rhabdomyomas.
- TSC should be suspected in infants with infantile spasms and ash-leaf spots.

16245

- Tuberous sclerosis complex should be suspected in a patient with refractory epilepsy, developmental delay, intellectual disability, and characteristic skin findings (eg, ash-leaf spots).
- Associated tumors include subependymal nodules and cardiac rhabdomyomas.

Von hippel lindau disease

18700

- Von Hippel-Lindau disease, an autosomal dominant condition caused by a mutation of a tumor suppressor gene, leads to various neoplasms, including CNS and retinal hemangioblastomas, pheochromocytomas, and renal cell carcinomas.

Wilson disease

2938

- Wilson disease presents with hepatic, neurologic, and psychiatric symptoms.
- The diagnosis is supported by low serum ceruloplasmin, increased urinary copper, and Kayser–Fleischer rings on slit-lamp examination.

Cerebrovascular disease

Amaurosis fugax

3528

- Amaurosis fugax is characterized by painless, rapid, and transient monocular vision loss.
- The description of a curtain descending over the visual field is highly suggestive of this diagnosis.
- The most common etiology is retinal ischemia due to atherosclerotic emboli originating from the ipsilateral carotid artery; therefore, patients with vascular risk factors should receive a duplex ultrasound of the neck.

Carotid artery stenosis

3529

- The surgical management of carotid atherosclerotic disease depends on whether it is asymptomatic or symptomatic, as well as the degree of stenosis.
- Patients with asymptomatic disease and stenosis <50% do not benefit from carotid revascularization.
- Instead, they are managed with intensive medical therapy (eg, antiplatelet agent, statin) and periodic carotid duplex ultrasonography to assess for progression of lesions.

4287

- All patients with carotid atherosclerotic disease should receive intensive medical management (ie, aspirin, statin, blood pressure control) and counseling on lifestyle changes to reduce future stroke risk.
- Carotid endarterectomy is also generally recommended for patients with symptomatic stenosis of 70%-99%.

Cavernous sinus thrombosis

3327

- Because the facial/ophthalmic venous system is valveless, uncontrolled infection of the skin, sinuses, and orbit can result in cavernous sinus thrombosis.
- This often presents with elevated intracranial pressure, bilateral periorbital edema, and deficits in the oculomotor (CN III), trochlear (CN IV), trigeminal (CN V), and abducens (CN VI) nerves.

Cranial nerve palsy

4408

- Ischemic oculomotor (CN III) palsy is commonly associated with poorly controlled diabetes mellitus and is characterized by damage to the inner somatic nerve fibers while sparing the more peripheral parasympathetic fibers.
- This manifests with ptosis, "down-and-out" gaze, diplopia, and normal pupillary response.

16619

- Non–pupil-sparing oculomotor nerve (CN III) palsies are concerning for aneurysmal compression.
- MR or CT angiography should be performed immediately.
- Pupil-sparing CN III palsies are most commonly caused by microvascular ischemia associated with diabetes mellitus, hypertension, and hyperlipidemia.

Giant cell arteritis

18576

- Giant cell arteritis is characterized by new headache and elevated inflammatory markers in patients age ≥50; complications include permanent vision loss.
- Definitive diagnosis can be obtained with temporal artery biopsy; however, to reduce the risk of blindness, treatment with systemic glucocorticoids should not be delayed while awaiting biopsy results.

4443

- Glucocorticoid-induced myopathy is a complication of chronic corticosteroid use.
- It is characterized by painless proximal muscle weakness, which is more prominent in the lower extremities.
- There is no muscle inflammation or tenderness, and creatine kinase level and erythrocyte sedimentation rate are normal.
- Glucocorticoid-induced myopathy slowly improves once the offending medication is discontinued.

3164

- Giant cell arteritis (GCA, or temporal arteritis) presents with headache and systemic symptoms (eg, fatigue, fever) in patients age ≥50.
- Potential complications include anterior ischemic optic neuropathy, which can lead to blindness.
- Threatened (eg, vision change) or confirmed vision loss in GCA requires high-dose intravenous glucocorticoids.

Hemorrhagic stroke

14528

- Intracerebral hemorrhage (ICH) typically presents with progressive headache, nausea/vomiting, and altered mental status over a period of minutes to hours.
- ICH in young patients is commonly due to arteriovenous malformation, which may also present with recurrent headache, seizure, or focal neurologic deficits.

4702

- In patients with suspected stroke, noncontrast CT scan of the head should be performed emergently to differentiate hemorrhagic versus ischemic stroke and guide further management.

19304

- Management of hemorrhagic stroke focuses on preventing further bleeding (eg, blood pressure control, anticoagulant reversal) and maintaining normal intracranial pressure.
- Hypertension is treated with a reversible and titratable antihypertensive (eg, intravenous nicardipine).

16284

- Cerebellar hemorrhage presents with progressive headache, nausea, and vomiting; in addition, vertigo, ipsilateral truncal (cerebellar vermis) or limb (cerebellar hemispheres) ataxia, dysarthria, and nystagmus may occur. CT scan demonstrates a posterior fossa hyperdensity.
- Urgent surgical decompression is indicated in patients with signs of neurologic deterioration or radiologic evidence of a hemorrhage >3 cm, brainstem compression, or obstructive hydrocephalus.

3737

- Patients who develop serious bleeding (eg, intracerebral hemorrhage) due to excess anticoagulation with warfarin should have their anticoagulation immediately reversed with intravenous vitamin K and prothrombin complex concentrate to reduce the risk of death and permanent disability.

12214

- Massive cerebral edema and/or hemorrhagic transformation, with rapid neurologic deterioration, can occur following a large, hemispheric ischemic stroke.
- Noncontrast CT scan of the head should be performed emergently to determine the extent of edema and/or hemorrhage and guide management.

3060

- Patients with hemorrhagic stroke initially tend to have focal symptoms that rapidly progress to signs of elevated intracranial pressure.
- Urgent noncontrast CT scan of the head is performed to confirm the hemorrhage.

18571

- Hemorrhagic stroke presents with sudden headache, focal neurologic deficits, and altered mental status.
- Risk factors in children include vascular malformations (eg, arteriovenous malformation [AVM]).
- Head CT scan of a ruptured AVM typically shows an intraparenchymal bleed (ie, hyperdense fluid collection with irregular margins).

20194

- Patients with intracerebral hemorrhage can have rapid clinical deterioration, especially when brain herniation is present.
- When a change in clinical status occurs, the ABCs (**A**irway, **B**reathing, **C**irculation) should be reassessed because intubation and mechanical ventilation may be necessary (eg, airway protection).

3537

- Cerebral amyloid angiopathy is associated with Alzheimer dementia and occurs due to the beta-amyloid deposition in the walls of small to medium cerebral arteries that leads to increased fragility of the vessels.
- The most common manifestation is a spontaneous, lobar intracranial hemorrhage.

2672

- The most common cause of spontaneous hypertensive intraparenchymal hemorrhage impacting the basal ganglia is hypertensive vasculopathy.
- It can also lead to hemorrhagic stroke involving the cerebellar nuclei, thalamus, and pons.

3728

- Strokes due to intracerebral hemorrhage often have focal neurologic deficits that progressively worsen over minutes to hours, followed by features of increased intracranial pressure.
- The basal ganglia (putamen) are common sites, and deficits often include contralateral hemiparesis, contralateral sensory loss, and conjugate gaze deviation toward the side of the lesion.

Ischemic stroke

4022

- An anterior cerebral artery stroke is characterized by contralateral motor or sensory deficits, which are more pronounced in the lower than in the upper limb.
- In addition, urinary incontinence can be seen due to disruption of the frontal micturition center.

4206

- Damage to the nondominant (right) parietal lobe can lead to hemineglect, which is characterized by unawareness of the left side of the world and/or body and impact sensory, motor, and conceptual functioning.

3738

- Broca aphasia is characterized by slow, nonfluent, effortful speech with intact language comprehension.
- Broca area is located adjacent to the primary motor cortex in the dominant frontal lobe and supplied by the superior division of the middle cerebral artery.

21302

- For patients with ischemic stroke, eligibility for thrombolysis and mechanical thrombectomy is considered independently.
- Depending on the time of presentation (<4.5 hr for thrombolysis, <24 hr for mechanical thrombectomy) and the presence or absence of specific contraindications, patients may be eligible for one, both, or neither of these therapies.

3879

- Transient ischemic attack and acute ischemic stroke are most commonly due to atherosclerotic cerebrovascular disease.
- Patients usually have multiple risk factors, but hypertension typically is the most important.
- Risk factor management should prioritize lowering blood pressure because even mild reductions can diminish stroke risk.

4921

- Cardioembolism (eg, due to atrial fibrillation) is a frequent cause of embolic stroke.
- Deficits are classically maximal at onset (due to rapid occlusion of the vasculature) and often localize to multiple vascular territories.

105586

- Young children (eg, age <6) with stroke often have nonfocal symptoms (eg, headaches, seizures, lethargy), in addition to classic focal neurologic deficits.
- Urgent MRI/MR angiography should be obtained to confirm the diagnosis and determine eligibility for potential reperfusion therapies.

4269

- A lateral medullary infarct (Wallenberg syndrome) leads to vertigo/nystagmus (vestibular nucleus), loss of pain/temperature sensation on the ipsilateral face (trigeminal nucleus) and contralateral body

(spinothalamic tract), bulbar weakness (lower cranial nerves), and ipsilateral Horner syndrome (descending sympathetic).

107696

- Patients with multiple sclerosis (MS) are at increased risk of ischemic stroke.
- Stroke symptoms can often be differentiated from symptoms of MS by the time course (hyperacute), localization to an arterial distribution, presence of cortical signs, or signs of increased intracranial pressure.

4482

- Lacunar strokes occur due to microatheroma formation and lipohyalinosis in the small penetrating arteries of the brain.
- They often affect the internal capsule and result in pure motor hemiparesis. Hypertension is the most important risk factor.

3712

- Patients with ischemic stroke who are not eligible for thrombolytic therapy or mechanical thrombectomy should receive antiplatelet agents (eg, aspirin, clopidogrel) to decrease the risk of early recurrent ischemic stroke.

17492

- Vertigo accompanied by loss of sensation over the ipsilateral face and contralateral body is concerning for a posterior circulation stroke that can be precipitated by vertebral artery dissection after minor neck trauma.

4203

- Lacunar stroke of the posterolateral thalamus typically presents with sudden-onset contralateral sensory loss involving all sensory modalities (ie, pure sensory stroke).
- Weeks to months later, patients can develop thalamic pain syndrome characterized by severe paroxysmal burning pain over the affected area that is exacerbated by light touch (allodynia).

4657

- Thrombolytic therapy (eg, intravenous alteplase) improves neurologic outcomes in patients with acute ischemic stroke when given within 4.5 hours of symptom onset.
- Before thrombolytics are administered, a noncontrast CT scan of the head should be performed to rule out hemorrhagic stroke, and the patient should be screened for other contraindications to therapy.

22066

- Sudden-onset, persistent vertigo accompanied by other neurologic symptoms (eg, dysmetria, ataxia, headache) is concerning for posterior circulation ischemia (eg, cerebellar stroke).
- Thrombolysis with intravenous alteplase should be administered to eligible patients with suspected ischemic stroke, even if classic stroke symptoms (eg, weakness, aphasia) are absent.

4268

- A stroke causing hemiparesis can usually be further localized based on the associated symptoms.
- A pure motor stroke with no cortical signs or cranial nerve deficits is likely due to a lesion in the contralateral internal capsule.

22064

- Ischemic stroke patients who are seen within 24 hours of symptom onset should have CT angiography of the head and neck performed to determine whether there is a large vessel occlusion amenable to treatment with mechanical thrombectomy.
- This occurs irrespective of thrombolysis being performed.

Mechanical valve

15833

- Prosthetic valve thrombosis (PVT) is a potential complication of both mechanical and bioprosthetic valves.
- Patients can have a new murmur, heart failure due to valvular obstruction (stenosis) or regurgitation, or a thromboembolic event (eg, transient ischemic attack).
- Suspected PVT should be promptly evaluated with echocardiography.

Retinal artery occlusion

2861

- Central retinal artery occlusion is a monocular painless acute vision loss most commonly caused by an embolized atherosclerotic plaque from the ipsilateral carotid artery.

Subarachnoid hemorrhage

18516

- Subarachnoid hemorrhage typically presents with a sudden-onset, severe headache that may be accompanied by vomiting, neck stiffness, fever, and loss of consciousness.
- CT scan of the head is the best initial diagnostic test.

3622

- Nontraumatic subarachnoid hemorrhage is most commonly caused by ruptured saccular (berry) aneurysm.
- It typically presents with thunderclap headache and signs/symptoms of meningeal irritation (eg, nuchal rigidity, photophobia, nausea).
- Noncontrast CT scan of the head usually reveals acute bleeding around the brainstem and basal cisterns.

18417

- Subarachnoid hemorrhage results in sudden-onset, severe headache with nausea and/or vomiting and other signs of meningeal irritation.
- Patients should be evaluated with urgent noncontrast CT scan of the head; if results are negative and a high suspicion for SAH remains, lumbar puncture should be performed.

18570

- Subarachnoid hemorrhage presents with a sudden-onset, severe headache.
- Lumbar puncture is warranted if clinical suspicion remains high despite a normal CT scan of the head.
- Characteristic findings include an elevated opening pressure, xanthochromia, and an elevated red blood cell count that does not decline with successive samples.

18517

- Isolated cranial nerve palsies can indicate a ruptured or enlarging saccular aneurysm with active or impending subarachnoid hemorrhage.
- Isolated oculomotor nerve palsy can indicate an aneurysm of the posterior communicating artery.

2678

- Delayed cerebral ischemia due to vasospasm is a major cause of delayed morbidity and mortality in subarachnoid hemorrhage.
- The risk can be reduced with initiation of nimodipine.

Subdural hematoma

4394

- Elderly patients are at higher risk for subdural hematoma due to increased fall risk and cerebral atrophy.
- Chronic subdural hematoma often presents insidiously weeks after the initial injury with headache, somnolence, confusion, and focal neurologic deficits.

Subclavian steal syndrome

12393

- Subclavian steal syndrome occurs due to stenosis or occlusion of the proximal subclavian artery, leading to reversal of blood flow in the ipsilateral vertebral artery.
- Patients are often asymptomatic but may have symptoms of upper extremity ischemia (eg, pain, fatigue, paresthesias) or vertebrobasilar insufficiency (eg, dizziness, ataxia, dysequilibrium) that are worsened by upper extremity exercise.

Transient ischemic attack

12180

- Neurological deficits related to an acute demyelinating plaque of multiple sclerosis usually last for days to weeks whereas transient ischemic attack (TIA) symptoms usually last for <24 hours.
- The initial treatment of TIA is modifying risk factors, starting aspirin and statin, and improving blood pressure control.

Vascular dementia

21186

- A single cerebral infarct may lead to vascular dementia (VaD) if it damages key areas involved in cognition (eg, medial frontal lobes, medial temporal lobes, thalamus).
- Patients with VaD due to a strategic infarction typically experience an abrupt decline in cognitive functioning.

Vertigo

12215

- Central vertigo presents with nonfatigable nystagmus, which cannot be suppressed by visual fixation. Head imaging should be performed to evaluate for cerebellar stroke or hemorrhage in patients who have persistent, new-onset central vertigo and vascular risk factors, neurologic signs/symptoms, or accompanying headache.

CNS Infections

Brain abscess

4248

- Direct spread of bacteria from otitis media or mastoiditis can cause a temporal brain abscess.
- The presentation can include severe headache, morning vomiting, fever, and focal neurologic deficits. Diagnosis is confirmed by visualization of a ring-enhancing lesion on CT scan or MRI of the brain.

4513

- A single brain abscess usually results from direct extension of an adjacent infection (eg, otitis media, sinusitis, dental infection).
- Viridans streptococcus and Staphylococcus aureus are the 2 most commonly isolated organisms. Headache, fever, focal neurologic deficits, and seizure may be present.
- Brain imaging (CT scan, MRI) typically reveals a single ring-enhancing lesion with central necrosis.

Cryptococcal infections

16124

- Cryptococcal meningoencephalitis is common in patients with advanced AIDS and generally manifests with signs of elevated intracranial pressure (ICP), including progressive headaches, nausea/vomiting, and confusion.
- In a minority of patients, elevated ICP compresses the 6th cranial nerve and results in diplopia and lateral gaze palsy.
- The diagnosis is generally made using lumbar puncture with CSF analysis and India ink stain or capsular polysaccharide antigen testing.

19821

- Patients with severe immunosuppression (eg, AIDS, solid-organ transplantation, immunosuppressant medications) are at increased risk for cryptococcal meningitis.
- Most cases are marked by subacute, progressive headache, vomiting, fever, and nuchal rigidity.
- Cerebrospinal fluid typically shows lymphocytosis with a mildly elevated white blood cell count, mild protein elevation, and low glucose.

Cytomegalovirus

12291

- Sensorineural hearing loss is the most common sequela of congenital cytomegalovirus (CMV).
- To reduce the risk of hearing loss, antiviral therapy should be given to neonates with symptomatic CMV (eg, hepatosplenomegaly, jaundice, periventricular calcifications).

Herpes Zoster

18582

- The classic triad of herpes zoster oticus (ie, Ramsay Hunt syndrome) includes severe ear pain, ipsilateral facial paralysis, and a vesicular rash in the external auditory canal.

16204

- Postherpetic neuralgia is characterized by burning pain and hyperesthesia lasting >4 months following acute zoster.
- The risk is greatest in those with advanced age, severe initial pain, or severe rash.
- First-line treatment includes anticonvulsants (eg, gabapentin) and tricyclic antidepressants (eg, amitriptyline).

HSV infection

4139

- Herpes simplex virus (HSV) encephalitis mainly affects the temporal lobe of the brain and may present acutely (<1 week duration) with focal neurological findings.
- The characteristic cerebrospinal fluid findings are lymphocytic pleocytosis, an increased number of erythrocytes, and elevated protein.
- HSV polymerase chain reaction analysis is the gold standard for diagnosis.

Meningitis

2442

- Hearing loss is a common sequela of Streptococcus pneumoniae meningitis due to inflammatory damage to the cochlea and/or labyrinth.
- Audiologic testing should be performed as soon as possible to identify hearing loss and improve outcomes if hearing rehabilitation is needed.

15232

- Among patients with meningitis, a viral etiology is suggested when cerebrospinal fluid analysis reveals a mildly elevated white blood cell count and protein level and normal glucose concentration.
- Mumps virus is capable of causing viral meningitis in unvaccinated individuals.

Septic emboli

4483

- Infective endocarditis often causes several weeks of nonspecific symptoms, complications related to septic emboli (eg, brain abscess), and new heart murmur.
- Initial workup includes 3 sets of blood cultures and echocardiography.

Syphilis

20552

- Neurosyphilis can occur at any point during syphilis infection.
- Meningovascular syphilis is usually marked by days or weeks of meningeal symptoms followed by acute ischemic infarction.
- Diagnosis is usually made with brain imaging and cerebrospinal fluid analysis, which is usually positive for VDRL.

12125

- Late neurosyphilis manifests years after untreated Treponema pallidum infection with general paresis (progressive dementia) or tabes dorsalis (sensory ataxia, lancinating pains, reduced/absent deep tendon reflexes).
- Patients often have Argyll Robertson pupils (normal pupillary constriction with accommodation but not with light).
- Intravenous penicillin is the treatment of choice.

Tuberculosis

19421

- Tuberculous meningitis is typically marked by subacute symptoms of meningeal irritation (eg, vomiting, headache, nuchal rigidity) and imaging findings of basilar meningeal enhancement, hydrocephalus, and stroke due to vasculitis.
- Risk is increased with immunocompromise (eg, HIV).

19419

- Tuberculous meningitis is generally marked by subacute symptoms of meningeal irritation with or without cranial nerve palsy and stroke.
- Cerebrospinal fluid analysis generally reveals lymphocytosis, moderate increase in white blood cells, mildly elevated protein, low glucose, and elevated adenosine deaminase.

Tick Paralysis

2289

- Tick-borne paralysis is characterized by rapidly progressive ascending paralysis (which may be asymmetrical), absence of fever and sensory abnormalities, and normal CSF examination.
- Ticks must feed for 4-7 days and are typically found on patients after meticulous searching.
- Removal of the tick results in spontaneous improvement in most patients.

Demyelinating diseases

Multiple sclerosis

107715

- Transverse myelitis (motor weakness with upper motor neuron signs, sensory deficits with a localizing spinal sensory level, and Lhermitte sign) is a common initial presentation of multiple sclerosis, which is an autoimmune disease that results in focal demyelination of the CNS.

2285

- Internuclear ophthalmoplegia is a disorder of conjugate horizontal gaze that results from damage to the medial longitudinal fasciculus (MLF).
- The affected eye (ipsilateral to the lesion) is unable to adduct, and the contralateral eye abducts with nystagmus.
- Multiple sclerosis plaques have a propensity to impact the MLF, either unilaterally or bilaterally.

15402

- Multiple sclerosis is an immune-mediated demyelinating disease of the central nervous system.
- The disease is most common in women age <50.
- Risk factors include vitamin D deficiency, geographic location, genetic predispositions (HLA-DRB1), and smoking.

2677

- A multiple sclerosis plaque in the upper thoracic spinal cord can result in paraplegia, bladder incontinence, and absent sensation below the level of the lesion.

3446

- Multiple sclerosis presents with neurologic deficits disseminated in both space and time.
- Initial presentations may not be disseminated in time, but further work-up and/or follow-up can lead to a confirmed diagnosis.

19661

- Multiple sclerosis can initially present (or often worsens) in the postpartum period.
- When multiple sclerosis is suspected, MRI of the brain showing demyelinating plaques disseminated in time and space can help confirm the diagnosis.

4381

- Pronator drift is a relatively sensitive and specific sign for upper motor neuron or pyramidal (corticospinal) tract disease affecting the upper extremities.
- It is performed by having the patient outstretch the arms with the palms up and eyes closed.
- In patients with upper motor neuron lesions, the affected arm drifts downward and the palm turns (pronates) toward the floor.

3643

- Acute exacerbations of multiple sclerosis with disabling neurologic symptoms (eg, motor, vision changes) are typically treated with high-dose glucocorticoids (eg, intravenous methylprednisolone).
- Plasmapheresis should be considered in patients who are refractory to corticosteroids.

107713

- Central vertigo often presents with nystagmus that is vertical, not fatigable, has no latency period, and does not change with head position.
- Postural instability is common, and patients often have other symptoms (eg, constitutional, other neurologic signs).
- Head imaging should be performed to evaluated for possible causes, including stroke or multiple sclerosis.

4641

- Definitive diagnosis of multiple sclerosis requires objective evidence of inflammatory demyelination disseminated in space and time.
- Acute attacks are treated with high-dose glucocorticoids.

3431

- Multiple sclerosis presents with neurologic deficits disseminated in space and time.
- Common initial presentations include transverse myelitis, optic neuritis, internuclear ophthalmoplegia, and cerebellar dysfunction.
- These initial presentations may not be disseminated in time, but further work-up and follow-up can confirm the diagnosis.

3462

- Trigeminal neuralgia is characterized by recurrent and sudden-onset severe, stabbing pain along branches of the trigeminal nerve (CN V).
- Although commonly thought to be due to vascular compression of the trigeminal nerve root, it can also be secondary to multiple sclerosis, likely due to inflammatory plaque formation affecting the trigeminal nuclei and/or nerve roots.

3644

- Multiple sclerosis (MS) presents with neurologic deficits disseminated in time and space.
- When MS is suspected but clinical examination or MRI is not classic, a lumbar puncture should be performed for cerebrospinal fluid analysis, which shows oligoclonal bands in >85% of patients.
- Opening pressure, protein, and cell count are generally normal.

Optic neuritis

- 2856
- Optic neuritis is usually characterized by the acute onset of monocular vision loss with central scotoma, afferent pupillary defect, changes in color perception, and pain with eye movement. There is a strong association between optic neuritis and multiple sclerosis.

Disorders of peripheral nerves and muscles

ALS

19242

- Amyotrophic lateral sclerosis can present with bulbar symptoms (eg, dysphagia, dysarthria) with examination revealing both upper motor neuron signs (eg, pathologic jaw jerk reflex) and lower motor neuron signs (eg, tongue fasciculations and atrophy).

Acute intermittent porphyria

6913

- Acute intermittent porphyria is a disorder of the heme synthesis pathway that results in reduced activity of porphobilinogen deaminase.
- Patients experience discrete attacks of abdominal pain, peripheral neuropathy with proximal muscle weakness, autonomic dysfunction, and neuropsychiatric manifestations.
- Urobilinogen levels are typically elevated, and elevated urinary porphyrin levels confirm the diagnosis.

GBS

4271

- Guillain-Barré syndrome most commonly presents with ascending weakness and areflexia, and can progress to involve respiratory and bulbar muscles.
- Serial pulmonary function testing (eg, spirometry) of forced vital capacity and negative inspiratory force is indicated to monitor for impending respiratory failure.

2290

- Guillain-Barré syndrome (GBS) is characterized by ascending weakness, bulbar symptoms (eg, dysarthria) and respiratory compromise after antecedent illness such as respiratory or gastrointestinal infection (especially *Campylobacter jejuni*).
- Cerebrospinal fluid analysis shows albuminocytologic dissociation.
- Treatment of GBS includes intravenous immunoglobulin or plasmapheresis.

3630

- Diarrheal illness (eg, Campylobacter jejuni infection) is the most frequent precipitant of Guillain-Barré syndrome (GBS), an acute demyelinating polyneuropathy characterized by ascending weakness and hyporeflexia.
- Other common findings of GBS include paresthesia, neuropathic pain, and autonomic dysfunction.

19489

- Guillain-Barré syndrome (GBS), an immune-mediated polyneuropathy, can be associated with early HIV infection.
- The clinical presentation and diagnostic workup of HIV-associated GBS is the same as non–HIV-associated GBS; nerve conduction studies (eg, showing demyelination) can help confirm the diagnosis.

3664

- Guillain-Barré syndrome typically presents with ascending weakness and diminished/absent deep tendon reflexes following a recent gastrointestinal or respiratory infection.
- The pathogenesis of this syndrome is the immune-mediated demyelination of peripheral nerve fibers.

19805

- Guillain-Barré syndrome presents with rapidly progressive ascending motor weakness due to immune-mediated demyelination of peripheral nerve fibers.
- Because it primarily affects peripheral nerves, MRI of the spinal cord is often normal.

16196

- Miller Fisher syndrome (MFS) is a variant of Guillain-Barré syndrome, a group of immune-mediated polyneuropathies that are caused by molecular mimicry.
- MFS is characterized by ophthalmoplegia, ataxia, and areflexia; strength is often preserved.
- It is highly associated with anti-GQ1b antibody.

4465

- Guillain-Barré syndrome is an acute or subacute ascending flaccid paralysis.
- Cerebrospinal fluid analysis shows an elevated protein level with normal cell count (albuminocytologic dissociation).

Bell's palsy

21510

- Slowly progressive facial weakness associated with hearing loss and facial twitching is concerning for a tumor at the cerebellopontine angle (eg, acoustic neuroma).

21518

- Bell palsy, a peripheral neuropathy involving the facial nerve (CN VII), presents with acute, progressive, unilateral facial weakness.
- In patients with a classic presentation, no further diagnostic workup is needed, and many other causes of facial weakness can be eliminated through a thorough history and physical examination.

14156

- Bell palsy (CN VII peripheral neuropathy) presents with acute, progressive, unilateral facial weakness involving the upper and lower face.
- A prodrome of auricular pain may be present.
- Therapy is with glucocorticoids, possibly in combination with acyclovir or valacyclovir.

Benign paroxysmal positional vertigo

16583

- Benign paroxysmal positional vertigo causes brief (<1 min) episodes of vertigo triggered by changes in head position.
- Triggering an episode by using provocative head positioning maneuvers (eg, Dix-Hallpike) is diagnostic.

Botulism

14459

- Wound botulism occurs when Clostridium botulinum spores contaminate a puncture injury (eg, intravenous needle), germinate, and generate neurotoxin in vivo.
- Manifestations include symmetric, descending neurologic deficits (eg, cranial nerve palsy), respiratory compromise, and autonomic dysfunction.
- In contrast to foodborne and infant botulism, fever and leukocytosis may be present.
- Urgent treatment with equine botulinum antitoxin is required and should not be delayed for diagnostic evaluation.

12120

- Foodborne botulism is caused by the ingestion of food contaminated with botulinum toxin and leads to bilateral cranial neuropathies and symmetric descending muscle weakness.
- Antitoxin treatment should be administered even before confirmatory diagnostic testing.

CIDP

17500

- Chronic inflammatory demyelinating polyneuropathy causes immune-mediated destruction of myelin in a non–length dependent manner; this results in progressive weakness of both proximal and distal muscles, sometimes with distal sensory loss.

4914

- Essential tremor can impair activities of daily living.
- First-line treatment includes propranolol, which exerts its effects due to peripheral beta-adrenergic receptor blockade, and primidone, a centrally acting anticonvulsant.

Copper deficiency

16772

- Copper deficiency typically occurs in patients with a history of gastric surgery (eg, bariatric), chronic malabsorption (eg, inflammatory bowel disease), or excessive zinc ingestion.
- Symptoms include slowly progressive myeloneuropathy similar to that of vitamin B12 deficiency (eg, distal extremity paresthesia, numbness, sensory ataxia), anemia, hair fragility, skin depigmentation, hepatosplenomegaly, edema, and osteoporosis.

Chorea

13756

- Sydenham chorea, a sequela of group A Streptococcus (GAS) infection, is the primary cause of chorea (ie, abnormal, jerky movements that disappear during sleep) in children.
- Hypotonia and behavioral changes are commonly seen, and evaluation includes GAS testing with throat culture and antistreptolysin O and antideoxyribonuclease titers.

Dystonia

4399

- Dystonia is a neurologic movement disorder characterized by sustained, involuntary muscle contractions that often result in abnormal, sometimes painful posturing.
- Many patients can have temporary relief with the use of a sensory trick (eg, touching the face).
- Associated tremor is common.

Diaphragmatic paralysis

18895

- Botulism presents with symmetric, descending motor paresis that can progress to involve the diaphragm, causing hypoventilation and respiratory acidosis.

Eaton lambert myasthenic syndrome

3837

- Lambert-Eaton myasthenic syndrome is frequently associated with an underlying malignancy (eg, small cell lung cancer).
- It is caused by autoantibodies directed against the voltage-gated calcium channels in the presynaptic motor nerve terminal leading to symmetric proximal muscle weakness with depressed deep tendon reflexes.

Femoral neuropathy

4293

- The femoral nerve is responsible for knee extension and hip flexion and provides sensation to the anterior thigh and medial leg.
- It is vulnerable to injury from pelvic fracture, hip dislocation, or iliacus hematoma and can suffer iatrogenic injury during prolonged maintenance of the dorsal lithotomy position (eg, hip/pelvic surgery, childbirth) or vascular procedures involving the femoral artery or vein.

Footdrop

12243

- Common fibular (peroneal) neuropathy can present with weakness on foot and toe dorsiflexion, along with impaired sensation over the lateral shin, dorsal foot, and web space between the first and second toes.
- Compression of the nerve at the fibular head for short periods (eg, sitting with legs crossed) can cause transient symptoms.

2634

- Myasthenia gravis is an autoimmune disease usually caused by autoantibodies directed against acetylcholine receptors; it presents with fatigable muscle weakness most commonly affecting the eyes and bulbar muscles.
- Thymic abnormalities (eg, hyperplasia, thymomas) are extremely common; chest imaging is indicated for diagnosis and surgical planning.

Hypokalemia

4393

- Hypokalemia is a common electrolyte abnormality that causes weakness, fatigue, and muscle cramps.
- When severe, it can lead to paralysis and arrhythmia.
- The ECG may show U waves, flat and broad T waves, and premature ventricular beats.

Median nerve injury

16491

- Carpal tunnel syndrome is common in patients with hypothyroidism and is frequently bilateral.
- Hypothyroidism causes soft tissue thickening and mucinous infiltration, which can lead to compression of the median nerve within the carpal tunnel.

16496

- Carpal tunnel syndrome is the most common mononeuropathy in patients with end-stage renal disease on dialysis.
- It is characterized by pain and paresthesia in the lateral hand; symptoms typically worsen during dialysis and are more severe in the arm with vascular access.

18962

- Because the median nerve lies directly adjacent to the brachial artery in the distal upper arm, it can be injured (eg, laceration, compression) as a result of brachial artery cannulation.

Myasthenia gravis

12028

- Symptoms of myasthenia gravis can be precipitated by surgery and the use of neuromuscular blocking agents.
- The diagnosis can be supported with a bedside test in which an ice pack is applied over the eyelids for several minutes.
- This inhibits the breakdown of acetylcholine at the neuromuscular junction, leading to an improvement in ptosis.

18453

- Myasthenia gravis is characterized by fluctuating, fatigable weakness affecting the eyes (eg, diplopia, ptosis), bulbar muscles (eg, dysphonia, difficulty chewing), and/or extremities (proximal > distal).
- The diagnosis is typically confirmed with acetylcholine receptor antibody assay.

3722

- Myasthenia gravis is characterized by fluctuating, fatigable weakness that involves the eyes, bulbar muscles, and/or extremities.
- Pyridostigmine, a long-acting oral acetylcholinesterase inhibitor, is the initial drug of choice because it increases the availability of acetylcholine at the neuromuscular junction.

3809

- Myasthenic crisis is a life-threatening complication of myasthenia gravis characterized by severe respiratory muscle weakness leading to respiratory failure.
- It may be precipitated by infection, surgery, and certain medications and heralded by increasing generalized or bulbar muscle weakness.

3727

- Management of myasthenic crisis with respiratory failure consists of endotracheal intubation followed by treatment with plasmapheresis (or intravenous immunoglobulins) and corticosteroids.

2667

- Pregnant and postpartum patients are at increased risk for developing myasthenia gravis, which is characterized by fluctuating, fatigable muscle weakness that worsens with repetitive motions and improves with rest.
- It commonly affects the eyes (eg, diplopia, blurred vision, ptosis), bulbar muscles (eg, dysphagia), and/or extremities (proximal > distal).

Myotonic Dystrophy

17088

- Myotonic dystrophy is a multisystem genetic disorder that classically causes muscle weakness and myotonia.
- Sleep disturbances, including excessive daytime sleepiness, are a common yet often underrecognized feature.
- The childhood phenotype (age of onset <10) often presents with intellectual impairment and behavioral difficulties.

Peripheral neuropathy

18492

- HIV neuropathy is a distal, symmetrical polyneuropathy that typically begins with numbness/tingling/pain in the feet and progresses proximally.
- Older patients with long-standing, poorly controlled HIV are at greatest risk.
- Antiretroviral treatment reduces the risk of progression.
- Gabapentin is first-line for symptomatic management.

16197

- Alcoholic neuropathy is a toxic neuropathy that typically results in a symmetric distal polyneuropathy (ie, stocking and glove pattern) characterized by paresthesia, burning pain, and ataxia.
- Loss of distal deep tendon reflexes (eg, ankle) and light touch and vibratory sense are common.

19267

- Progressive sensory loss in a distal, symmetric, stocking-glove distribution is characteristic of a length-dependent axonal polyneuropathy.
- If it occurs acutely, it is likely to be related to a toxin or medication such as metronidazole.

Peroneal neuropathy

4427

- Foot drop is caused by injury to nerves supplying the foot dorsiflexors (eg, L5 radiculopathy, common fibular [peroneal] neuropathy).
- Patients must flex the hip and knee to avoid dragging the foot, which results in a unilateral high-stepping, or steppage, gait.

Polyarteritis nodosa

18608

- Polyarteritis nodosa often causes mononeuritis multiplex, a neuropathy of ≥2 noncontiguous peripheral nerves.
- Patients also frequently have skin, renal, and gastrointestinal manifestations; the lungs are generally spared.

Restless legs syndrome

4687

- Restless legs syndrome is characterized by an uncomfortable urge to move the legs that is temporarily relieved by movement.
- Alpha-2-delta calcium channel ligands (eg, gabapentin) are recommended as first-line therapy.
- Dopamine agonists (eg, pramipexole, ropinirole) are also effective but can cause a paradoxical worsening of symptoms with long-term use.

Sarcoidosis

16472

- Sarcoidosis is a systemic granulomatous disorder that can involve the nervous system, presenting as facial nerve palsy.
- Other common extrapulmonary manifestations of sarcoidosis include lymphadenopathy, hypercalcemia, and parotid gland swelling.

Shoulder dislocation

4604

- Acute shoulder pain after forceful abduction and external rotation at the glenohumeral joint suggests an anterior shoulder dislocation, which may cause injury to the axillary nerve.
- The axillary nerve innervates the teres minor and deltoid muscles, and injury can result in weakened shoulder abduction and decreased sensation over the lateral shoulder.

Spasticity

19797

- Multiple sclerosis may result in disabling spasticity.
- Baclofen, an agonist at the GABA-B receptor, and tizanidine, an alpha-2 adrenergic agonist, are the most commonly used medications for reducing muscle tone and painful muscle spasms.

Syncope

3963

- Vasovagal syncope is a type of reflex syncope typically preceded by a trigger (eg, strong emotion) and a prodrome (eg, pallor, light-headedness).
- A neurally mediated reflex response (cardioinhibitory and vasodepressor) leads to brief (eg, <1-2 min) loss of consciousness, followed by rapid and complete recovery.

Tetanus

12265

- Tetanus is a presynaptic neuromuscular junction disorder caused by the Clostridium tetani toxin, which blocks the release of the inhibitory neurotransmitters glycine and GABA across the synaptic cleft, leading to painful muscle spasms and trismus (lockjaw).
- It is seen almost exclusively in patients who are unvaccinated or incompletely immunized.

Tremor

4913

- Essential tremor most often presents as a tremor of the hands that is suppressed at rest, exacerbated by outstretched arms, and more pronounced at the end of goal-directed movements.
- It is not associated with other neurologic symptoms.

Trigeminal neuralgia

3460

- Trigeminal neuralgia is caused by compression of the trigeminal nerve (CN V) root as it enters the pons, usually by an abnormal vessel loop.
- This leads to atrophy and demyelination of the nerve and causes short paroxysms of neuropathic pain.

Ulnar nerve neuropathy

4170

- The ulnar nerve is vulnerable to injury where it runs posterior to the medial epicondyle at the elbow.
- Features include decreased sensation in the medial hand and medial 1½ digits, decreased grip strength, and weakness of intrinsic hand muscles.

Vitamin b12 deficiency

16313

- Long-term (≥5 years) metformin therapy can cause vitamin B12 deficiency due to impaired absorption of vitamin B12 in the terminal ileum.
- This can often (>40% of patients) present with neurologic findings (eg, paresthesia, sensory ataxia, neuropsychiatric changes) and normal hematologic studies.

10499

- Vitamin B12 deficiency is common in those with chronic malabsorption (eg, Crohn disease) and often presents with macrocytic anemia and neurologic findings (eg, paresthesia, subacute combined degeneration).
- A vitamin B12 level is the initial test of choice, but serum methylmalonic acid and homocysteine testing should be ordered in borderline or inconclusive cases.

Hydrocephalus

16273

- Newborns who do not receive intramuscular vitamin K are at risk of vitamin K deficiency bleeding, which presents with easy bruising or bleeding, including intracranial hemorrhage (ICH).
- ICH can cause obstructive hydrocephalus, leading to a bulging fontanelle, upward gaze impairment, and signs of increased intracranial pressure (eg, irritability, vomiting, bradycardia, hypertension).

18664

- Macrocephaly is a head circumference >97th percentile.
- The condition is most likely benign (eg, familial) in a patient with normal development and normal examination (eg, no syndromic features, no signs of increased intracranial pressure).
- Management is reassurance and observation.

18665

- Myelomeningocele is an open neural tube defect in which the meninges and spinal cord protrude through the skin.
- It is often associated with a Chiari II malformation (ie, inferior displacement of the medulla and cerebellum through the foramen magnum), as well as obstructive hydrocephalus.

107416

- Ventriculoperitoneal shunt malfunction, which most commonly occurs within 6 months of placement, leads to obstruction of cerebrospinal fluid flow and recurrence of hydrocephalus.
- Signs and symptoms are due to increased intracranial pressure (eg, headache, lethargy, vomiting) and white matter tract compression (eg, incontinence, weakness, hyperreflexia).

4871

- Hydrocephalus should be suspected in infants with rapidly increasing head circumference and signs of increased intracranial pressure (eg, full fontanelle).
- The first step in evaluation is ultrasonography (if fontanelle is open) of the head or MRI.

15238

- Signs of increased intracranial pressure (eg, irritability, vomiting, lethargy) in patients with a ventriculoperitoneal shunt should prompt urgent evaluation for shunt complications (eg, infection, malfunction), including neurologic evaluation, CT scan of the head, and shunt series x-rays.

18663

- Hydrocephalus should be suspected in an infant or young child with a rapidly enlarging head circumference crossing multiple growth percentiles.
- Signs/symptoms of increased intracranial pressure are more commonly seen once the anterior fontanelle closes and include irritability, developmental delay, hypertension/bradycardia, and papilledema.

17517

- Choroid plexus papilloma is a benign intraventricular mass that causes increased production of cerebrospinal fluid, leading to ventriculomegaly and hydrocephalus.
- In infants, hydrocephalus presents with enlarging head circumference and signs of increased intracranial pressure.

Headache

Hemorrhagic stroke

19403

- Cocaine use can precipitate intracranial hemorrhage and should be suspected when stroke occurs in a subcortical location and/or in young patients with associated sympathetic activation or an absence of typical risk factors.

Cluster headache

3935

- Cluster headaches manifest with excruciating, unilateral periorbital pain, as well as restlessness and autonomic symptoms.
- The most effective abortive treatment is 100% oxygen via face mask.

4253

- Cluster headaches usually present with acute, unilateral, severe periorbital pain that awakens patients from sleep.
- These headaches are often accompanied by autonomic symptoms (eg, tearing, ipsilateral ptosis, miosis).
- The most effective abortive therapy is 100% oxygen.

114669

- Primary stabbing headache presents with sharp, jabbing pain that is extremely short and has a very focal location.
- This benign condition can typically be managed with reassurance, but if symptoms are bothersome, indomethacin can be used as preventive therapy.

Cerebral venous thrombosis

19620

- Cerebral vein and venous sinus thrombosis is associated with prothrombotic conditions (eg, postpartum) and has a highly variable presentation that may include headache, increased intracranial pressure, seizures, and/or stroke.
- Diagnosis is confirmed by visualizing obstructed venous flow on MR venography of the brain.

107472

- Cerebral vein and venous sinus thrombosis has a variable presentation due to unique pathophysiology that may include headache, increased intracranial pressure, seizures, and/or focal deficits from venous stroke/hemorrhage.
- Diagnosis is confirmed by visualizing obstructed venous/sinus flow on MR venography.

Glaucoma

4367

- Angle-closure glaucoma is characterized by narrowing of the anterior chamber angle leading to decreased aqueous outflow and elevated intraocular pressure.
- It presents with headache, eye pain, nausea, and decreased visual acuity.
- Examination findings include conjunctival injection, corneal edema, palpable firmness of the eyeball, and a fixed, mid-dilated pupil.
- The diagnosis is confirmed by gonioscopy and/or tonometry.

Migraine

17490

- Migraine is an episodic neurologic disorder that results in severe, unilateral, throbbing headaches often associated with photophobia, phonophobia, and nausea/vomiting; they may be accompanied by aura (focal, reversible neurologic symptoms that precede or accompany the headaches).
- Estrogen-containing contraceptives are contraindicated in patients with migraine with aura due to the increased risk for ischemic stroke.

19006

- Migraines are classically episodic, unilateral, throbbing headaches commonly triggered by stress, fasting, and/or dehydration.
- Acute management includes simple analgesia (eg, acetaminophen) and a triptan (eg, sumatriptan) in refractory cases.

3672

- Migraines in children can cause unilateral or bifrontal pain with photophobia, phonophobia, nausea, and/or vomiting.
- Visual auras (eg, flashing lights, scintillating scotoma) can precede the headache.
- Diagnosis is clinical, and first-line treatment includes acetaminophen or nonsteroidal anti-inflammatory drugs.

18968

- Preventive migraine therapy may benefit patients with severe migraine headaches (eg, ≥4 times per month, significant impairment).
- During pregnancy, first-line prevention is with beta blockers (eg, propranolol, metoprolol).

3619

- Patients with frequent and/or severe migraine episodes that lead to disabling symptoms preventing regular activities (eg, missing work) should be offered a daily preventive medication to reduce the frequency, duration, and intensity of headaches.
- First-line preventive medications include beta blockers (eg, metoprolol), tricyclic antidepressants (eg, amitriptyline), and anticonvulsants (eg, valproate).

18976

- Classic migraine presents with a unilateral and pulsatile headache, phonophobia, photophobia, nausea, and/or vomiting.
- Common triggers include sleep deprivation and menses.
- Imaging and laboratory evaluation are not warranted for patients with classic symptoms and normal neurologic examination findings.

Idiopathic Intracranial Hypertension

3081

- Intracranial hypertension presents with headache, vision changes, papilledema, and/or abducens nerve (CN VI) palsy.
- Growth hormone, tetracyclines, and excessive vitamin A and its derivatives (eg, isotretinoin) can cause increased intracranial pressure.
- Withdrawal of these medications leads to symptom resolution.

3637

- Idiopathic intracranial hypertension presents with positional headaches, vision changes, and papilledema with normal neuroimaging and an elevated opening pressure on lumbar puncture.
- If there is no immediate threat to vision, first-line treatment is weight loss and acetazolamide.

4900

- Increased intracranial pressure can lead to swelling of the optic nerve (CN II) head (ie, papilledema), which can manifest as transient vision loss with changes in head position.
- Enlargement of the optic nerve head also causes enlarged physiologic blind spots.

3044

- Idiopathic intracranial hypertension occurs most often in young, obese women and presents with headaches suggestive of a brain tumor, but with normal neuroimaging and elevated cerebrospinal fluid pressure.
- The most serious complication is blindness.
- Treatment includes weight loss and carbonic anhydrase inhibitors (eg, acetazolamide).
- Shunting or optic nerve (CN II) sheath fenestration may be performed in patients experiencing progressive visual defects despite therapy.

16457

- Idiopathic intracranial hypertension is most common in obese women of childbearing age and can present with positional headaches, pulsatile tinnitus, and papilledema.
- Diagnosis is with MRI of the brain followed by lumbar puncture.

107461

- Idiopathic intracranial hypertension presents with headaches, vision changes, and papilledema.
- After neuroimaging is performed, lumbar puncture demonstrating an elevated opening pressure (>250 mm H2O) confirms the diagnosis.

16456

- Although idiopathic intracranial hypertension typically presents in obese women of childbearing age, it can also be seen in nonobese children.
- Headaches may be less obvious, and vision abnormalities (eg, blurry vision, enlarged blind spot) are often predominant in the prepubertal population.
- Papilledema is the hallmark finding.

3162

- Idiopathic intracranial hypertension typically presents in young, obese women with headache, vision changes, papilledema, and sometimes abducens nerve (CN VI) palsy.
- Diagnosis is confirmed by lumbar puncture showing elevated opening pressure and normal cell counts.

Increased intracranial pressure

4708

- Headaches due to idiopathic intracranial hypertension can be difficult to distinguish clinically; further evaluation is often required, especially in obese women of childbearing age with progressively worsening symptoms.
- Normal neuroimaging with an increased opening pressure and normal composition of the cerebrospinal fluid on lumbar puncture are diagnostic of idiopathic intracranial hypertension.

12228

- Neuroimaging should be obtained in patients with headaches that present with warning features.
- These warning features include pain that is new or different in character, associated with neurologic deficits, or accompanied by signs of increased intracranial pressure (eg, nausea and vomiting, blurry vision).

Medication overuse headache

14457

- Medication-overuse headache is characterized by chronic, near-daily headaches (usually in the morning) in the setting of regular use of analgesics.
- It most commonly occurs in patients with a preexisting headache disorder.
- Management involves cessation of the analgesic, which may cause transiently increased headaches.

Temporomandibular disorders

18480

- Temporomandibular joint disorder can present with headaches, muscle spasms, jaw fatigue, facial pain.
- Physical examination can reveal tenderness of the muscles of mastication, as well as pain, crepitus, and/or audible clicks from the joint.

Tension headache

12241

- Tension-type headaches can occur due to stress and are usually bilateral, dull, and nonthrobbing without associated symptoms (eg, nausea, vomiting).

- In most cases, these headaches can be treated with simple analgesics, but prophylactic medication (eg, amitriptyline) should be considered when they are more disabling.

Miscellaneous

Alcohol withdrawal

18287

- Patients with alcohol use disorder are at risk for alcohol withdrawal with alcohol reduction/cessation.
- Manifestations begin within 6-24 hours and include anxiety, agitation, tremor, diaphoresis, and nausea.
- More severe cases are marked by delirium tremens (eg, autonomic instability, delirium), hallucinations, and/or seizures.
- The primary treatment is with benzodiazepines, which dampen CNS excitation.

Aminoglycoside

2837

- Aminoglycosides can be ototoxic to both the cochlea (resulting in sensorineural hearing loss) and the vestibular system (resulting in imbalance).

Anticholinergics

20443

- Patients treated with multiple anticholinergic medications (eg, elderly psychiatric patients) are at risk for chronic anticholinergic toxicity, which often presents with subtle and nonspecific findings not typical of acute anticholinergic toxicity.
- Removal of the anticholinergic medication leads to resolution of symptoms.

Acute kidney injury

10776

- Asterixis is a flapping movement of the hands that occurs when the wrist is extended with the arms outstretched.
- Common causes include hepatic encephalopathy, uremic encephalopathy, and hypercapnia.
- Treating the underlying cause will improve neurological status and resolve asterixis.

Air embolism

17007

- Vascular air embolism can occur due to decompression sickness following deep underwater diving.
- Venous air embolism can cause skin cyanosis, respiratory distress, and obstructive shock, whereas arterial air embolism can cause confusion, gait ataxia, dysarthria, and stroke.

Benzodiazepines

3383

- Benzodiazepines should be used with extreme caution in the elderly due to increased risk of cognitive impairment, falls, and paradoxical agitation.

Breath holding spells

4872

- Breath-holding spells are characterized by brief apnea and color change (ie, cyanosis or pallor) associated with an emotional trigger and followed by loss of consciousness.
- Parents should be reassured that these spells are typically benign with no long-term consequences.

Carbon monoxide poisoning

14195

- Acute carbon monoxide poisoning can occur due to inhalation of car exhaust in a closed space, often performed intentionally in a suicide attempt.
- Toxicity results from impaired delivery and usage of oxygen, leading to clinical manifestations of cerebral hypoxia (eg, headache, confusion, seizure, coma).
- Laboratory results may demonstrate lactic acidosis, and permanent hypoxic brain injury can occur.

Chorea

20307

- Sydenham chorea is an autoimmune complication of group A Streptococcus infection caused by molecular mimicry, in which antistreptococcal antibodies cross-react with neuronal antigens in the basal ganglia.
- Neuropsychiatric manifestations include chorea, milkmaid grip, hypotonia, emotional lability, and obsessive-compulsive behaviors.

Delirium

21328

- Delirium is characterized by acute-onset, fluctuating consciousness, most frequently seen in elderly hospitalized patients.
- It usually resolves with treatment.
- However, cognitive dysfunction can be persistent, and a single episode of delirium increases the risk of further cognitive decline.

21331

- Delirium is an acute, fluctuating disturbance in attention, thinking, and consciousness.
- Abnormal findings on initial evaluation guide further investigation (eg, chest x-ray due to fever, crackles, and leukocytosis) and treatment.
- If initial evaluation is negative or there is inadequate response to treatment, extended investigation is warranted.

4622

- Toxic-metabolic and infectious etiologies are the most common causes of delirium in a hospitalized patient.
- Patients with dementia have an increased risk of developing agitated delirium in the hospital.
- Typical and atypical antipsychotics are useful for treating acute agitation in elderly patients with dementia.
- Benzodiazepines are typically not recommended in this setting.

4700

- Wernicke encephalopathy is due to thiamine (vitamin B_1) deficiency and is most commonly seen in malnourished patients with underlying alcoholism.
- Features include encephalopathy, ocular dysfunction, and gait ataxia.
- Patients should be treated empirically with thiamine prior to or along with glucose administration.

12248

- Delirium is a reversible disorder characterized by agitation and confusion, most frequently seen in the elderly or those with underlying dementia.
- Determining the underlying cause, avoiding unnecessary medications, treating infections and metabolic disturbances, and encouraging regular activity during the day while minimizing disturbances at night can help patients recover more quickly.

21333

- Nonpharmacologic management of delirium symptoms is preferred whenever possible.
- Personal interaction and constant observation reduce the need for physical restraints and can be provided by family members or professional sitters.

4644

- Delirium is common but under-recognized.
- The Confusion Assessment Method improves delirium identification using its fundamental features of acute-onset, fluctuation, inattention, disorganized thinking, and altered consciousness.
- Initial evaluation is directed at the most common causes, but further assessment should follow any abnormal findings or negative initial evaluation.

Heat related illness

4703

- Heat stroke is characterized by core temperature ≥40 C (104 F) with CNS dysfunction (eg, altered mental status).
- It occurs most commonly in those exposed to hot/humid environments while performing extreme activities.
- Complications include rhabdomyolysis, disseminated intravascular coagulation, and end-organ dysfunction.

Hyponatremia

114914

- Chronic hyponatremia manifestations can range from subtle (eg, nausea, fatigue) to more pronounced (eg, encephalopathy, gait disturbances, seizures).
- Thiazides (eg, chlorthalidone) can cause hyponatremia, particularly in elderly women with a low BMI; patients are typically euvolemic.
- Initial management includes thiazide cessation.

Lead poisoning

12396

- Chronic lead toxicity presents in adults with neuropsychiatric, gastrointestinal, and general symptoms, including peripheral sensorimotor neuropathy, fatigue, abdominal pain, and constipation.
- It can lead to hypertension, nephropathy, hyperuricemia, and microcytic anemia with basophilic stippling seen on peripheral smear.
- Treatment for symptomatic patients involves chelation therapy with an agent such as calcium disodium EDTA.

12395

- Chronic lead toxicity in adults presents with neuropsychiatric, gastrointestinal, and general symptoms including peripheral sensorimotor neuropathy, fatigue, abdominal pain, and constipation.
- It can lead to hypertension, nephropathy, hyperuricemia, and microcytic anemia with basophilic stippling seen on the peripheral smear.
- Diagnosis hinges on a thorough history including potential sources of lead exposure.

Lithium

16272

- Chronic lithium toxicity most commonly results from reduced lithium clearance due to decreased renal perfusion (eg, dehydration).
- Patients typically develop neurologic symptoms including lethargy, confusion, ataxia, and tremor.
- Cardiac abnormalities (eg, QT interval prolongation, bradycardia) may also be present.

Mild cognitive impairment

21721

- Review of medications should be prioritized in evaluating new-onset or worsening cognitive impairment.
- Medications with anticholinergic properties are strongly associated with risk for cognitive impairment, particularly in the elderly.

Thiamine deficiency

3082

- Thiamine deficiency can cause Wernicke encephalopathy, which is characterized by encephalopathy, oculomotor dysfunction, and gait ataxia.
- This is generally seen in malnourished patients (eg, anorexia, chronic alcohol use) and may be induced iatrogenically by the administration of glucose without thiamine.

14369

- Wernicke encephalopathy is a neurologic complication of thiamine deficiency; it classically manifests as the triad of ataxia, encephalopathy, and oculomotor dysfunction.
- It is usually associated with long-standing heavy alcohol use; however, it may be caused by any disorder that causes chronic malnourishment (eg, short-gut syndrome).
- Treatment is with intravenous thiamine.

Tourette syndrome

20778

- Pharmacotherapy of Tourette syndrome consists of vesicular monoamine transporter type 2 (VMAT2) inhibitors, antipsychotic medications, and alpha-2 adrenergic receptor agonists.
- VMAT2 inhibitors are increasingly preferred as initial treatment over antipsychotics due to a more favorable adverse effect profile.

Tremor

19546

- Functional (psychogenic) tremors often present abruptly with significant disability.
- They often decrease with distraction, have changeable or inconsistent features, and are not consistent with known tremor syndromes.

4179

- Essential tremor most often presents as a tremor of the hands that is suppressed at rest, exacerbated by outstretched arms, and more pronounced at the end of goal-directed movements.
- It is often hereditary and can be associated with a head tremor, but it is not associated with other neurologic symptoms.
- The diagnosis is clinical.

Tricyclic antidepressants

12302

- Amitriptyline is commonly used for depression, insomnia, and pain disorders but is frequently associated with side effects, including orthostatic hypotension, lethargy, and anticholinergic symptoms (eg, dry mouth, constipation, urinary retention).
- Discontinuing the medication usually resolves these symptoms.

Neurodegenerative disorders and dementias

Alzheimer disease

16032

- The elderly are especially vulnerable to the cumulative adverse effect burden of polypharmacy.
- In particular, medications with anticholinergic properties are strongly associated with the risk for cognitive impairment, which may produce a medication-induced syndrome that mimics dementia.
- Review of medications is an important first step in evaluating a patient on a complex medication regimen with new-onset or worsening cognitive impairment.

3373

- Cholinesterase inhibitors such as donepezil, rivastigmine, and galantamine may provide moderate symptomatic relief of cognitive symptoms and temporarily improve functioning in Alzheimer disease; however, these medications have not been shown to alter the disease course.

4620

- Alzheimer disease, the most common type of dementia in the United States, is characterized by early and prominent memory impairment.
- The differential includes dementia with Lewy bodies, frontotemporal dementia, and vascular dementia, as well as nondementing syndromes such as normal pressure hydrocephalus.

19472

- Patients with a family history of Alzheimer disease are at increased risk of developing the disease but can mitigate risk by addressing modifiable risk factors.
- Aggressive treatment of cardiovascular risk factors (eg, hypertension, diabetes, obesity/physical inactivity), especially in mid-life, can help reduce risk.

12246

- The first step in evaluation of suspected mild cognitive impairment is to perform a standardized cognitive examination using a cognitive screening tool such as the Montreal Cognitive Assessment (MoCA).
- Patients with evidence of possible cognitive impairment on the MoCA should undergo a workup for potentially reversible causes, which includes depression screening, laboratory evaluation, and neuroimaging.

3058

- Alzheimer disease initially presents with memory impairment.
- Neuroimaging generally demonstrates temporal lobe atrophy, which is most prominent in the hippocampi and surrounding medial temporal lobes.

16082

- The initial workup of suspected dementia should include neuropsychological testing (eg, Montreal Cognitive Assessment), selected laboratory testing (complete blood count, complete metabolic panel, TSH, vitamin B12), and neuroimaging (eg, MRI).
- Patients in specific risk groups may warrant additional targeted testing.

3074

- Alzheimer disease, the most common cause of dementia, is characterized by insidious memory loss followed by behavioral changes.
- Psychotic symptoms (delusions, hallucinations) may develop later in the course of the disease.

ALS

14196

- Amyotrophic lateral sclerosis leads to degeneration of the upper motor neurons (eg, motor nuclei in the precentral gyrus and their projections in the corticospinal and corticobulbar tracts) and lower motor neurons (eg, cranial nerves, spinal nerves).
- Because any motor neuron can be involved, patients may have a variety of symptoms, including bulbar muscle weakness.
- Examination shows both upper motor neuron and lower motor neuron signs.

3771

- Amyotrophic lateral sclerosis presents with both upper and lower motor neuron signs; sensation is typically normal.
- Medical management includes riluzole (a glutamate inhibitor) and edaravone (an antioxidant), which improves survival and slows disease progression.

19217

- Amyotrophic lateral sclerosis leads to degeneration of cells in the anterior horn of the spinal cord.
- It is characterized by often asymmetric weakness with upper and lower motor neuron signs.
- Electrodiagnostic studies often show characteristics of both acute and chronic denervation.

19212

- Amyotrophic lateral sclerosis can cause respiratory insufficiency due to respiratory muscle weakness.
- Noninvasive positive-pressure ventilation prolongs survival and improves quality of life.

Cerebellar degeneration

12247

- Alcoholic cerebellar degeneration is caused by damage to the Purkinje cells of the cerebellar vermis.
- Manifestations typically include the slowly progressive onset of ambulation difficulties, including wide-based gait and postural instability.
- Truncal coordination (eg, tandem gait) is impaired, but limb coordination (eg, finger-nose test) is relatively intact.

Dementia with lewy bodies

4619

- Dementia with Lewy bodies causes cognitive fluctuations, visual hallucinations, and parkinsonism.
- Cognitive symptoms may precede, or appear along with, parkinsonian features.

Depression

2350

- Sleep disturbances are commonly seen in depression.
- New-onset insomnia in elderly patients who have associated symptoms of depression should raise concern for major depressive disorder.

4060

- Depression-related cognitive impairment may be so severe in elderly patients that they appear to have dementia.
- Psychotherapy and/or antidepressant medications are the treatment of choice for elderly patients with depression and frequently result in reversal of cognitive deficits.

3919

- In older patients, cognitive impairment caused by depression often mimics other neurocognitive disorders, such as Alzheimer disease.
- However, it is accompanied by other symptoms of depression and usually improves with treatment of the underlying depressive disorder.

Frontotemporal dementia

2281

- Frontotemporal dementia is a type of early-onset (age 50s-60s) dementia.
- It is characterized by early and progressive personality (apathetic, nonempathetic), behavior (impulsive, compulsive), and executive function abnormalities.

Glaucoma

4401

- Acute angle-closure glaucoma (ACG) is a vision-threatening condition that may be associated with certain medications (eg, anticholinergics for Parkinson disease) or arise spontaneously.
- Manifestations include the sudden onset of severe eye pain, nausea, vomiting, unilateral conjunctival injection, and a dilated pupil with poor light response.
- Untreated, patients with acute ACG can develop permanent vision loss within 2-5 hours of symptom onset.
- Urgent ophthalmologic consultation is required.

Huntington disease

15787

- Huntington disease is an autosomal dominant disease of CAG trinucleotide repeat expansion characterized by psychiatric symptoms, cognitive impairment, and chorea.
- It is associated with preferential degeneration of GABA-producing neurons in the caudate nucleus and putamen.

3631

- Huntington disease is an autosomal dominant, CAG trinucleotide repeat expansion disease presenting with psychiatric symptoms, cognitive impairment, and chorea.
- It is associated with preferential atrophy of the caudate nucleus and putamen.

Multiple system atrophy

4130

- Multiple system atrophy is a Parkinson-plus syndrome characterized by motor parkinsonism (eg, rigidity, bradykinesia, resting tremor) with early, significant autonomic dysfunction.
- Falls, cerebellar dysfunction, and pyramidal signs are also often present.

Normal pressure hydrocephalus

15950

- Normal pressure hydrocephalus (NPH) is characterized by ventriculomegaly with normal opening pressure on lumbar puncture.
- It classically presents with a triad of incontinence, cognitive impairment, and gait abnormalities; however, all symptoms may not be present in early disease.
- NPH can be idiopathic or occur secondary to neurologic insults (eg, subarachnoid hemorrhage, trauma, meningitis) that result in scarring of the arachnoid granulations responsible for cerebrospinal fluid resorption.

15951

- Normal pressure hydrocephalus is a potentially reversible cause of dementia characterized by cognitive decline, gait impairment, and urinary incontinence.
- Neuroimaging demonstrates ventriculomegaly out of proportion to the sulci.

4652

- Normal pressure hydrocephalus is characterized by abnormal gait, urinary incontinence, and cognitive impairment; however, only gait dysfunction is required for diagnosis.
- Neuroimaging demonstrates ventriculomegaly out of proportion to cerebral atrophy.
- The diagnosis is confirmed with high-volume lumbar puncture demonstrating improvement of gait with cerebrospinal fluid removal.
- Definitive treatment is ventricular shunt placement.

4651

- Normal pressure hydrocephalus is characterized by gait abnormalities, cognitive impairment, and urinary incontinence that often presents as urgency in early disease.
- It is thought to result from decreased CSF absorption with increased ventricular compliance, allowing for ventricular enlargement without chronically increasing intracranial pressure.

3923

- Normal pressure hydrocephalus is characterized by the symptom triad of gait disturbance, dementia, and urinary incontinence; upper motor neuron signs may also occur.
- Neuroimaging demonstrates enlargement of the ventricles out of proportion to the sulci.

Parkinson disease

21687

- Depression is a common complication of Parkinson disease (PD) and is often undertreated because the two conditions have overlapping symptoms (eg, psychomotor slowing [bradykinesia], blunted affect [masked facies]).
- Patients with depression can be treated with antidepressant medication, psychotherapy, or both.

4322

- Parkinsonism often leads to a hypokinetic gait characterized by difficulty initiating steps, a narrow base, shuffling or festinating strides, asymmetrically reduced arm movement, and en bloc turning.

14962

- Parkinson disease dementia (PDD) is characterized by executive and visuospatial dysfunction with relatively mild memory impairment at first.
- PDD may be distinguished from dementia with Lewy bodies by the timing of symptom onset: If parkinsonism predates cognitive impairment by >1 year, PDD should be diagnosed.

12486

- Major depressive disorder may occur in up to 20% of patients with Parkinson disease (PD).
- Although the diagnosis of major depression may be difficult due to overlapping symptoms with PD, the presence of depressed mood, anhedonia, hopelessness, and/or suicidality often suggests major depression.

18528

- Autonomic dysfunction is common with Parkinson disease and can lead to neurogenic orthostatic hypotension, with patients experiencing lightheadedness or syncope on standing.
- The disorder is typically recognized by a ≥20 mm Hg drop in systolic blood pressure with standing and an absence of the expected increase in heart rate.

106786

- Parkinson disease can present with bulbar symptoms, including aspiration and hypophonia (soft voice). Olfactory dysfunction is also common.
- Examination often shows rigidity, stooped posture, and typical gait changes (eg, slowness, shuffling).

3718

- Parkinson disease (PD) should be suspected in patients with an asymmetric resting tremor that is associated with rigidity or bradykinesia.
- Trihexyphenidyl is an anticholinergic medication sometimes used in the treatment of PD, generally in younger patients where tremor is the primary symptom.

4563

- The cardinal signs of Parkinson disease are rest tremor, rigidity, and bradykinesia.
- At least 2 of these on physical examination are grounds for a clinical diagnosis.

4400

- Tremor is often the presenting symptom of Parkinson disease.
- It is usually a resting, pill-rolling tremor that asymmetrically impacts the hands.
- It improves with voluntary actions, worsens when patients are distracted, and is not impacted by caffeine or alcohol.

Prion disease

2990

- Creutzfeldt-Jakob disease is a fatal prion disease that presents with rapidly progressive dementia, myoclonus, mood changes, and sleep disturbances.
- There is no effective disease-modifying treatment.
- Patients rapidly decline until death, which occurs less than 12 months after diagnosis.

15786

- Creutzfeldt-Jakob disease is a fatal prion disease that presents with rapidly progressive dementia, myoclonus, mood changes, and sleep disturbances.
- Cerebrospinal fluid findings are typically noninflammatory and often include high levels of 14-3-3 protein.

3374

- Creutzfeldt-Jakob disease is a rapidly fatal prion disease that presents with rapidly progressive dementia, myoclonus, mood symptoms, and sleep disturbances.
- MRI of the brain often reveals hyperintensity of the cerebral cortex (ie, cortical ribboning) and the putamen and caudate head (ie, hockey stick sign).

Vascular dementia

15783

- Vascular dementia is caused by cerebral ischemia or infarction and commonly presents with executive dysfunction, focal neurologic findings, and subcortical symptoms/signs.
- It typically follows a progressively declining course.

10448

- Vascular dementia following stroke presents as sudden or stepwise decline in executive function that interferes with activities of daily living.
- Patients often have focal neurologic findings on examination (eg, hemiparesis, pronator drift, Romberg sign) due to prior (potentially unrecognized) strokes.

10348

- Vascular dementia classically presents as a sudden or stepwise decline in executive function with mild memory loss.
- Patients can have focal neurologic deficits on examination, and neuroimaging classically demonstrates multiple small infarctions.

Amaurosis fugax

6996

- Amaurosis fugax is painless, sudden, and transient (<10 minutes) monocular vision loss that most commonly results from retinal artery emboli originating from an ipsilateral carotid artery atherosclerotic plaque.

Multiple sclerosis

6981

- Multiple sclerosis is an inflammatory demyelinating disorder that typically presents in women age 15-50 with neurologic deficits disseminated in space and time.
- Patients frequently develop optic neuritis characterized by transient painful monocular vision loss that improves after several weeks.
- Heat exposure may exacerbate symptoms (Uhthoff phenomenon).

Antipsychotics

4899

- Antipsychotics cause hyperprolactinemia by blocking dopamine activity in the tuberoinfundibular pathway.
- Clinical effects of hyperprolactinemia include amenorrhea, galactorrhea, gynecomastia, and sexual dysfunction.

Hearing loss

13790

- Chronic, excessive noise exposure can lead to sensorineural hearing loss due to the irreversible death of hair cells in the cochlea.
- Hearing screening programs are often mandated in high-risk occupations (eg, manufacturing, transportation).

11996

- Presbycusis is common in elderly patients and presents with bilateral, symmetric, sensorineural hearing loss.

Seizures and epilepsy

Anoxic brain injury

16265

- Myoclonus is characterized by sudden, involuntary muscle contraction or relaxation that results in movement of limbs or joints.
- Posthypoxic myoclonus is often seen after cardiac arrest; it occurs while patients are unconscious and is characterized by generalized, symmetric myoclonus that involves the axial, limb, and facial muscles.
- Myoclonus status epilepticus is a marker of poor prognosis.

Seizures

2680

- Seizure should be considered in any patient with an episode of sudden loss of consciousness followed by a postictal state of sleepiness and confusion.
- Other common findings include perioral cyanosis, evidence of tongue biting, and urinary incontinence.

2280

- Focal seizures originate in a single cerebral hemisphere and can spread to involve both hemispheres, causing impairment of awareness (with or without automatisms) and inability to respond to external stimuli.
- In contrast to absence seizures, focal seizures are associated with a postictal phase and are not provoked by hyperventilation.

4378

- Seizures are characterized by sudden loss of consciousness, loss of postural tone, and a postictal state with delayed return to baseline neurologic functioning.
- Tongue lacerations can occur with tonic-clonic movements.

19890

- Patients who remain unresponsive after generalized convulsive status epilepticus require electroencephalography to differentiate between sedation due to benzodiazepines and persistent nonconvulsive status epilepticus.

110558

- Neuroimaging should be obtained after an unprovoked, first seizure in adults.
- Seizure can be the presenting symptom of brain metastases.

4155

- Todd paralysis is a transient, focal weakness or paralysis (eg, hemiplegia) that occurs in the postictal period after a focal-onset (± secondary generalization) seizure.
- Symptoms are self-limited and usually resolve within 36 hours.

2681

- Absence seizures typically occur in children age 4-10 and are characterized by <20-second episodes of impaired concentration (eg, pause, blank stare) with preserved postural tone.
- Electroencephalography shows generalized 3-Hz spike-wave activity, and ethosuximide is the first-line treatment.

2671

- CT scan of the head without contrast is the initial imaging study of choice in patients with first-time seizure to exclude acute neurologic conditions (eg, intracranial or epidural bleed) that might require urgent intervention.
- MRI is more sensitive than CT scan in identifying most structural causes of epilepsy and is the neuroimaging modality of choice in elective situations.

11982

- The initial diagnostic work-up of a first-time seizure in an adult should include basic blood tests (serum electrolytes, glucose, calcium, magnesium, complete blood count, and renal and liver function tests) and a toxicology screen to evaluate for metabolic and toxic causes.
- Unprovoked seizures generally require further evaluation with MRI and electroencephalography.

14370

- Generalized convulsive status epilepticus is defined as a single seizure lasting ≥5 minutes or ≥2 seizures in which the patient does not completely regain consciousness.
- Intravenous benzodiazepines (eg, lorazepam, diazepam) should be administered for seizure termination.
- In addition, a nonbenzodiazepine antiepileptic medication should be administered to prevent seizure recurrence; recommended medications include levetiracetam, fosphenytoin, or valproic acid.

4089

- Patients with status epilepticus (ie, a single seizure for ≥5 minutes or ≥2 seizures without interictal return to neurological baseline) are at increased risk for developing permanent injury (ie, lasting neurologic deficits) due to excitatory cytotoxicity.
- This occurs predominantly in the cortex because it is the site of seizure origin (ie, cortical laminar necrosis).

12044

- Phenytoin is an antiepileptic drug with known teratogenic effects (eg, fetal hydantoin syndrome).
- Women of childbearing age who have a low risk for seizure recurrence may safely discontinue phenytoin if considering becoming pregnant; however, the medication should be slowly tapered because rapid withdrawal may result in seizure recurrence.

Antiepileptics

16291

- Phenytoin is highly protein-bound and metabolized hepatically by the cytochrome p450 system; medications that inhibit cytochrome p450 (eg, trimethoprim-sulfamethoxazole, fluconazole) or that displace phenytoin from plasma proteins (eg, valproic acid) increase the risk of drug toxicity.
- Common symptoms of acute phenytoin toxicity include horizontal nystagmus, ataxia, and nausea/vomiting, and significant toxicity can result in altered mental status, coma, and death.

Conversion disorder

15124

- Psychogenic nonepileptic seizure (PNES) is a type of conversion disorder commonly misdiagnosed as a seizure disorder.
- Features suggestive of PNES include forced eye closure, side-to-side head or body movements, memory recall of the event, and lack of postictal confusion.
- The gold standard for diagnosis of PNES is video-electroencephalogram of an event demonstrating lack of epileptiform activity.

Febrile seizure

4841

- Simple febrile seizures are typically benign and occur in children age 6 months to 5 years.
- They are generalized, last <15 minutes, and do not recur within 24 hours.
- Diagnostic testing is unnecessary, and management is with supportive care (eg, antipyretics) and reassurance.

Breath holding spells

16073

- Pallid breath-holding spells are triggered by fear or pain from minor trauma and are characterized by pallor/diaphoresis with brief loss of consciousness that may be followed by sleepiness/confusion.
- Diagnosis is clinical and requires no testing.

Sleep disorders

REM sleep behavior disorder

15146

- REM sleep behavior disorder (RBD) is characterized by dream enactment during REM sleep atonia.
- Most patients with idiopathic RBD eventually develop a disorder of alpha-synuclein neurodegeneration, most commonly Parkinson disease.

Restless legs syndrome

20487

- Restless legs syndrome is characterized by an urge to move the legs with discomfort that is relieved by movement.
- The risk is increased in patients with certain chronic neurologic disorders (eg, multiple sclerosis).
- If pharmacotherapy is required for patients with frequent or severe symptoms, alpha-2-delta calcium channel ligands (eg, pregabalin, gabapentin) are recommended.

20485

- Restless legs syndrome (RLS) is characterized by an urge to move the legs and discomfort that is temporarily relieved by movement.
- It occurs primarily in the evenings and at night and can interfere with the quality and duration of sleep.
- RLS is diagnosed clinically; however, serum iron studies should be obtained, and iron supplementation should be started in patients with low or low-normal ferritin levels.

20486

- Restless legs syndrome can occur during pregnancy and is characterized by an urge to move the legs accompanied by dysesthesia that is worsened by inactivity and improved with movement.
- Symptoms are worse in the evening or at night and may impair sleep.

Narcolepsy

3470

- Narcolepsy is characterized by excessive daytime sleepiness, cataplexy, hypnagogic/hypnopompic hallucinations, and sleep paralysis.
- Treatment includes sleep hygiene, scheduled naps, and avoidance of alcohol and drugs that cause drowsiness.
- When medications are needed to decrease daytime somnolence, wakefulness-promoting agents such as modafinil are preferred.

Spinal cord disorders

Anterior cord syndrome

4204

- Thoracic aortic aneurysm repair can cause spinal cord ischemia, especially of the anterior cord.
- Anterior cord syndrome typically presents with distal, bilateral flaccid paralysis; loss of pain/temperature and crude touch sensation; and urinary retention.

16079

- Descending (type B) aortic dissections can be associated with interruption of blood flow to the anterior spinal cord, leading to anterior spinal cord ischemia (particularly in the lower thoracic levels).
- Patients generally develop acute bladder paralysis, lower extremity paresis, crude touch/pain sensation loss, and diminished reflexes (initially).
- Vibrioception and deep touch sensation are generally preserved because these are carried in the posterior columns and receive some blood flow from the intact posterior spinal arteries.

3300

- Anterior cord syndrome usually occurs when there is injury to the anterior spinal artery from trauma.
- It is characterized by bilateral motor function loss at and below the level of the injury with diminished pain and temperature sensation bilaterally that begins 1-2 levels below the cord injury, whereas proprioception, vibratory sensation, and light touch are unaffected.

Brown-sequard hemi cord syndrome

4274

- Hemisection of the spinal cord can cause Brown-Séquard syndrome, which is characterized by the following:
- Ipsilateral hemiparesis and diminished proprioception, vibratory sensation, and light touch at the level of the spinal cord injury and below
- Contralateral diminished pain and temperature sensation 1-2 levels distal to the cord injury and below

Cauda equina syndrome

4392

- Cauda equina syndrome, a neurologic emergency, is a disorder marked by compression of ≥2 lumbosacral nerve roots.
- The diagnosis should be considered in patients with severe, radicular pain plus impaired motor/sensory/reflex activity in the lower extremities, bowel/bladder/sexual dysfunction, and/or saddle anesthesia.

12244

- Severe radicular pain involving one or both legs with accompanying saddle anesthesia and bladder dysfunction is concerning for cauda equina syndrome (CES).
- CES is commonly caused by a large lumbosacral disc herniation.
- It requires urgent MRI of the lumbosacral spine and, in most cases, urgent surgical decompression.

106481

- Acute displacement of the nucleus pulposus (ie, disc herniation) can compress the spinal nerve roots or spinal cord, resulting in acute neurologic deficits.
- Compression of the S1 nerve root can result in loss of Achilles reflex and lower extremity weakness.

Cervical spondylosis

13878

- Cervical radiculopathy is common in older individuals and usually arises after physical activity or trauma puts stress on the neck and leads to disc herniation or nerve root compression from underlying cervical spondylosis.
- Patients usually have pain in the neck radiating to the shoulder/arm, weakness in an upper extremity myotome, and sensory loss in an upper extremity dermatome.
- MRI of the cervical spine is the first test of choice.

4205

- Cervical radiculopathy often arises in older individuals due to underlying cervical spondylosis.
- Manifestations generally include neck pain that radiates to the shoulder/arm and unilateral upper extremity sensory, motor, and/or reflex abnormalities in a dermatomal distribution.
- Spine imaging generally shows abnormal facet joints, including the presence of sclerosis and osteophytes.

Central cord syndrome

3072

- Hyperextension injury, especially in elderly patients with cervical spine degenerative changes (ie, cervical spondylosis), can cause central cord syndrome.
- This classically causes loss of pain and temperature sensation in the upper extremities and disproportionate upper extremity weakness.

Cervical myelopathy

14697

- Cervical spondylotic myelopathy is common in older adults and generally presents with progressive neck pain, gait disturbances, lower motor neuron signs in the upper extremities (eg, weakness, diminished reflexes), and upper motor neuron signs in the lower extremities (eg, increased reflexes, spasticity).

12245

- The most common cause of cervical myelopathy in older adults is spondylosis, a degenerative spine disease that causes canal narrowing with spinal cord compression.
- Manifestations include progressive gait instability and weakness in the extremities.
- Examination usually shows lower motor neuron signs at the level of the lesion (arms) and upper neuron signs below the level of the lesion (legs).

19322

- Cervical myelopathy often causes both spinal cord and spinal nerve root compression, resulting in myelopathic symptoms (eg, upper motor neuron signs below the lesion) and radicular symptoms (eg, lower motor neuron signs, pain in a dermatomal/myotomal pattern).
- Lhermitte sign (electric shock–like pain with neck flexion) may occur.

Depression

19321

- Cervical radiculopathies occur due to spinal nerve root compression and typically causes neck pain associated upper extremity sensorimotor deficits that follow a dermatomal/myotomal pattern.
- Lateral flexion and rotation of the neck worsens compression of the nerve root, worsening pain and/or paresthesia.

Epidural abscess

4372

- Spinal epidural abscess should be suspected in patients with fever, focal back pain, and acute neurologic deficits (eg, leg weakness, urinary retention).
- Emergency MRI of the spine is the diagnostic study of choice.

3983

- Spinal epidural abscess often presents with several days/weeks of fever, malaise, and the following progressive neurologic symptoms: focal back pain → nerve root pain → motor weakness, sensory changes, and bowel/bladder dysregulation → paralysis.
- Suspected cases require urgent MRI spine; treatment generally includes surgical decompression and antibiotics. Epidural anesthesia is a common triggering event (due to direct inoculation).

19891

- Spinal epidural abscess can occur due to trauma, especially in immunocompromised patients.
- Manifestations include progressive worsening of pain, neurologic deficits, and systemic inflammatory signs (eg, fever, leukocytosis).

Neural tube defects

21399

- Closed spinal dysraphism can present with signs of tethered cord, including bladder dysfunction and neurologic deficits (eg, weakness, hyporeflexia, abnormal gait).
- Foot deformities (eg, pes cavus, hammer toe) are common.

21398

- Closed spinal dysraphism (spina bifida occulta) is due to failed fusion of the vertebral arches.
- Symptomatic newborns can have cutaneous manifestations (eg, lumbosacral hemangioma) on the lower back and/or neurologic deficits due to tethered cord syndrome.
- Neurologic manifestations include lower motor neuron signs (eg, weakness, hypotonia, hyporeflexia, sensory loss).

21400

- Closed spinal dysraphism (spina bifida occulta) should be suspected in a patient with new-onset urinary incontinence, particularly if lumbosacral cutaneous findings (eg, hair tuft) are present.
- Diagnosis is with MRI of the spine, and surgical detethering is indicated in symptomatic patients.

21401

- Chronic back pain associated with neurologic deficits (eg, hyporeflexia, weakness) and a gross lumbosacral abnormality (eg, lipoma) should raise concern for tethered spinal cord with underlying spinal dysraphism.

Penetrating neck trauma

19881

- High spinal cord injuries (above C5) can lead to hypercapnic respiratory failure due to diaphragmatic paralysis.
- Even in patients with unstable cervical spine injuries, orotracheal intubation (with manual stabilization of the cervical spine) should be performed.

Radiculopathy

19320

- Cervical radiculopathy is due to nerve root compression and typically presents with neck or arm pain associated with sensorimotor deficits; radiation of pain with neck movement may occur.
- The diagnosis is usually made clinically, and most patients improve with symptomatic treatment, including nonsteroidal anti-inflammatory drugs and avoidance of triggering activities.

18469

- Cervical radiculopathy is usually caused by compression of the nerve root due to disk herniation or spondylosis; manifestations depend on the involved nerve root but typically include neck pain, shoulder/arm pain, weakness, paresthesia, and diminished reflexes.
- Improvement of radicular symptoms when the hand is placed on the top of the head is both diagnostic and therapeutic.

19325

- L5 radiculopathy often presents with low back pain that radiates down the leg associated with sensory loss over the lateral thigh, calf, and dorsal foot, and weakness of great toe extension and foot dorsiflexion, inversion, and eversion.

Rheumatoid arthritis

16182

- Patients with rheumatoid arthritis are at risk for atlantoaxial instability; neck extension during intubation can result in subluxation with cord compression and cervical myelopathy.
- Symptoms of cervical myelopathy include a slowly progressive, spastic paraparesis involving the upper and lower extremities, hyperreflexia, sensory changes, and a positive Babinski sign.
- Hoffman sign may also be positive.

Spinal cord compression

4391

- Back pain, symmetric extremity weakness, and signs of upper motor neuron dysfunction (eg, hyperreflexia) are concerning for spinal cord compression, a neurologic emergency.
- MRI of the spine should be performed immediately to determine the cause of compression and direct treatment.

3560

- Acute spinal cord compression can present with loss of motor and sensory function, loss of rectal tone, and urinary retention.
- Management includes emergency surgical consultation, neuroimaging, and possibly intravenous glucocorticoids.

2679

- Diabetic neuropathy is a distal, symmetric polyneuropathy that can cause a "stocking and glove" pattern of sensory loss, impaired proprioception and vibratory sense, and diminished ankle reflexes; motor findings may be a late manifestation in severe disease.
- Hyperactive reflexes and an upgoing plantar reflex are inconsistent with diabetic neuropathy and are suggestive of an upper motor neuron process.

4691

- Neoplastic epidural spinal cord compression presents with worsening focal back pain, bilateral lower-extremity weakness, sensory loss, and gait ataxia.
- Bowel/bladder disturbances are late findings. In the acute phase of spinal cord injury, patients can develop spinal shock with absence of reflexes and flaccid paraplegia as a result.

Spinal cord injury

16022

- Spinal cord injury above T6 can be complicated by autonomic dysreflexia, in which noxious stimuli below the injury level trigger an unregulated sympathetic response, leading to severe hypertension.
- A compensatory parasympathetic response above the lesion typically causes bradycardia.
- Management includes removing noxious stimuli and treating the hypertension.

17426

- CT scan is the preferred test to screen for cervical spine injury.
- Indications include high-energy mechanism of injury or any of the following findings: neurologic deficit, spinal tenderness, altered mental status, intoxication, or distracting injury.

16089

- Acute spinal cord injury often manifests with loss of spinal cord function (eg, areflexia, anesthesia, paralysis, distended bladder) below the level of the lesion.
- Lesions that arise above T1 also often cause neurogenic shock due to interruption of the descending sympathetic fibers, which results in unopposed parasympathetic stimulation of the vessels (hypotension) and heart (bradycardia).
- Hypothermia is also common due to a lack of peripheral vasoconstriction.

18312

- Cervical facet dislocation typically occurs with forced flexion of the cervical spine (eg, falling onto a flexed neck); a single facet is usually dislocated and results in radiculopathy of the corresponding nerve root.
- The most commonly affected vertebral bodies are C5/C6, which lead to C6 radiculopathy, and C6/C7, which lead to C7 radiculopathy.
- Imaging demonstrates anterior subluxation of the vertebral bodies.

18658

- CT scan is the preferred test to screen for cervical spine injury.
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18659

- The presence of a single vertebral fracture in a patient with blunt trauma is an indication to image the entire spine.
- CT scan is the screening modality of choice because of its sensitivity and accuracy.

3784

- In patients with traumatic spinal cord injury, disruption of the autonomic tracts involved in bladder control can lead to urinary retention.
- Therefore, catheterization should be performed to prevent bladder distension and possible injury.

16088

- Central cord syndrome is common after whiplash-type injuries in older adults with underlying cervical spondylosis.
- Damage to the central cervical spinal cord causes upper extremity motor, sensory, and reflex abnormalities; sacral (eg, bowel/bladder) and lower extremity function is generally preserved.

Spinal stenosis

2316

- Lumbar spinal stenosis is most commonly caused by degenerative joint disease.
- The term "neuropathic claudication" is often used to describe lumbar stenosis.
- This refers to the exacerbation of leg symptoms with walking (similar to peripheral vascular disease [PVD]).
- However, unlike PVD, the symptoms are positional and remain while standing still.
- Pain is relieved by flexion of the spine.
- Diagnosis is made based on clinical history and classic findings on spinal MRI.

Syringomyelia

3724

- Syringomyelia is a disorder in which a fluid-filled cavity forms within the spinal cord (usually within the cervical and/or thoracic spine).
- Patients typically have dissociated sensory loss (ie, loss of pain/temperature sensation but not vibratory/proprioceptive sensation), often in a "cape" distribution, and may develop weakness that disproportionately affects the upper extremities.

4698

- Syringomyelia is characterized by a fluid-filled cavity (syrinx) in the spinal cord that compresses the surrounding tissue.
- It is most commonly seen in patients with Arnold-Chiari type 1 malformations or spinal cord injury.
- It typically affects the crossing fibers of the spinothalamic tract in the ventral white commissure (loss of pain and temperature sensation with preserved touch, vibration, and proprioception) and may affect motor fibers in the ventral horns (flaccid paralysis).
- The diagnosis is confirmed by MRI, and management usually requires surgical intervention.

Transverse Myelitis

16473

- Transverse myelitis is an immune-mediated disorder characterized by the infiltration of inflammatory cells into a segment of the spinal cord.
- Inflammation localizes to ≥1 contiguous spinal cord segments, leading to a rapidly progressive myelopathy characterized by motor weakness, autonomic dysfunction (eg, bowel/bladder dysfunction), and sensory deficits with a distinct sensory level.

19736

- Transverse myelitis is an immune-mediated, rapidly progressive myelopathy that typically causes motor and sensory deficits, a distinct sensory level, and focal T2 hyperintensity of the spinal cord on MRI.
- First-line management is the administration of high-dose intravenous glucocorticoids.

Traumatic brain injuries

Brain death

4159

- Brain death, an irreversible loss of all brain function, is a clinical diagnosis based on clinical criteria and neurologic examination.
- Essential findings include coma, absent cranial nerve reflexes and brain-originating motor responses, and apnea.
- Movements originating in the spinal cord (eg, deep tendon reflexes) may be present.

Brain herniation

4552

- Rapid hematoma (eg, epidural) expansion after head injury can abruptly increase intracranial pressure, compress the temporal lobe, and cause uncal herniation.
- The first sign is typically an ipsilateral fixed and dilated pupil due to oculomotor nerve (CN III) compression.
- Contralateral hemiparesis is often seen due to direct compression of the ipsilateral cerebral peduncle.

20496

- Central herniation occurs when the diencephalon and midbrain are displaced caudally due to increased intracranial pressure (eg, intracranial hemorrhage).
- This causes damage to the brainstem, which results in Cushing triad, unconsciousness, midsized and fixed pupils, and abnormal posturing (decorticate vs decerebrate).

20493

- Subfalcine herniation, a type of brain herniation, occurs when the cingulate gyrus is displaced under the falx cerebri.
- This typically does not cause pupillary involvement but may cause ipsilateral anterior cerebral artery compression that leads to contralateral leg weakness.

Cervical artery dissection

4117

- Posterior oropharyngeal injuries can result in internal carotid artery dissection or thrombus formation, which can present with hemiplegia, facial droop, and aphasia.

16169

- Internal carotid artery dissection is a common cause of stroke in young patients and can occur spontaneously or after mild trauma or illness.
- It is typically characterized by partial Horner syndrome (ptosis and miosis without anhidrosis), unilateral headache and neck pain, and cerebral ischemia (transient ischemic attack, stroke).

Child abuse

3396

- Abusive head trauma is the most common cause of death from child abuse.
- Repetitive acceleration-deceleration forces cause shearing of the subdural bridging veins and vitreoretinal traction, resulting in subdural and retinal hemorrhages.

Concussion

18429

- Concussion is caused by rapid rotational acceleration of the brain during head trauma.
- Diagnosis is based on clinical findings of neurologic disturbance (eg, headache, noise sensitivity, emotional changes) without evidence of structural intracranial injury.
- Treatment is physical and cognitive rest followed by a gradual return to activity.

18426

- Concussion, or mild traumatic brain injury, is characterized by neurologic symptoms (eg, headache, difficulty concentrating) without structural intracranial injury.
- Initial treatment is with cognitive and physical rest for 24-48 hours, followed by a gradual return to play with slowly increasing physical intensity.

18412

- Concussions are characterized by a transient neurologic disturbance (eg, disorientation, amnesia) that results from mild traumatic brain injury (TBI); headache, dizziness, slurred or delayed speech, and incoordination are also common.
- After a concussion, average-risk adults should rest for ≥24 hours before gradually increasing activity level each 24-hour period they remain asymptomatic, potentially returning to full contact sports in 1 week.

18425

- Imaging should be avoided in children with minor head trauma and no high-risk features for intracranial injury (ie, altered mental status, loss of consciousness, severe mechanism of injury, vomiting or severe headache, signs of basilar skull fracture).

18284

- Concussion is a form of mild traumatic brain injury resulting in transient impairment of normal neuronal function.
- Typical symptoms include headache, disorientation, dizziness, and/or amnesia associated with abnormalities in coordination, speech, attention, or emotions.

3218

- Head CT scan without contrast is indicated for minor head trauma with high-risk features for intracranial injury (ie, altered mental status, loss of consciousness, severe mechanism of injury, vomiting or severe headache, signs of a basilar skull fracture).
- Observation for 4-6 hours may be an alternative option if mental status is normal and there are no signs of a basilar skull fracture.

Epidural hematoma

4923

- Epidural hematoma is most common in young adults and occurs as a result of traumatic head injury, which leads to tearing of the middle meningeal artery and bleeding between the dura mater and skull.
- CT scan of the head typically reveals a hyperdense biconvex lesion that does not cross suture lines.
- Symptomatic patients require emergent neurosurgical hematoma evacuation.

16480

- Epidural hematomas occur due to tearing of the middle meningeal artery and typically occur with skull fracture.
- Although patients classically present with loss of consciousness followed by a lucid interval, many initially remain alert.
- However, hematoma expansion results in neurologic decompensation with signs of elevated intracranial pressure (eg, headache, nausea/vomiting, altered mental status) within minutes to hours.

3297

- Epidural hematoma occurs when blood accumulates between the cranium and the dura mater after traumatic head injury.
- Patients classically have a brief loss of consciousness followed by a lucid interval.
- Rapid hematoma expansion can lead to elevated intracranial pressure and uncal herniation.

16481

- Spinal epidural hematoma is a potential complication of neuraxial anesthesia (eg, epidural block), lumbar puncture, or spinal surgery and is more common in older adults taking antithrombotic medications.
- Manifestations include slowly progressive motor and sensory dysfunction and localized back pain; bowel and bladder dysfunction may occur. Management includes an urgent MRI and neurosurgical decompression.

Orbital fracture

16718

- An orbital floor fracture can result in entrapment of the inferior rectus muscle, which presents with vertical diplopia and restriction of upward eye movement.

Post concussion syndrome

18456

- Postconcussion syndrome is characterized by prolonged (>4 weeks) concussion symptoms (eg, headache, sleep disturbance).
- Management is symptomatic care because most patients improve within 3 months.

11557

- Traumatic brain injury (TBI) can lead to postconcussion syndrome, which presents with physical, cognitive, and/or emotional symptoms.
- These symptoms typically resolve with symptomatic treatment within a few months following TBI.

Subdural hematoma

4153

- Subdural hematoma results from the rupture of bridging veins, most commonly from head trauma.
- Risk factors include advanced age and chronic alcoholism (due to brain atrophy), as well as anticoagulant use.
- On non-contrast head CT scan, acute subdural hematoma appears as a crescent-shaped hyperdensity that crosses suture lines.

Transverse myelitis

19740

- Osmotic therapy (eg, hypertonic saline, mannitol) is part of the initial treatment of elevated intracranial pressure in patients with traumatic brain injury.
- It creates an osmolar gradient that draws water out of the edematous brain tissue, thereby reducing parenchymal volume and overall intracranial pressure.

Traumatic brain injury

19024

- Meningiomas often occur at dural reflections and can cause headaches and progressive neurologic defects that localize to the brain area compressed by the mass.

3226

- Meningiomas often occur at dural reflections and can cause headaches and progressive neurologic defects that localize to the brain area compressed by the mass.

3299

- Diffuse axonal injury (DAI) often causes severe neurologic impairment in the absence of major CT scan findings.
- MRI is more sensitive for DAI and may reveal minute punctate hemorrhages in the white matter and blurring of the gray-white interface.

18942

- Meningiomas often occur at dural reflections and can cause headaches and progressive neurologic defects that localize to the brain area compressed by the mass.

18943

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Tumors of the nervous system

CNS lymphoma

2276

- Patients with advanced AIDS are at increased risk for malignancy.
- Primary CNS lymphoma is an AIDS-defining malignancy associated with the expression of Epstein-Barr virus oncogenes, which can often be identified in the cerebrospinal fluid.
- MRI of the brain generally shows a solitary, irregular, nonhomogenous ring-enhancing mass.

Brain tumors

3658

- In children, low-grade astrocytomas are the most common type of central nervous system tumor.
- New-onset seizures may be the initial presentation of a tumor originating in the cerebral cortex.

3725

- A glioblastoma often presents with headaches and signs of increased intracranial pressure.
- Characteristic imaging findings include a butterfly-shaped tumor (due to spread under the falx cerebri), serpiginous peripheral contrast enhancement, and central necrosis.

17518

- Craniopharyngiomas are benign, slow-growing, calcified tumors within the suprasellar region.
- Tumor compression of the optic chiasm can result in bitemporal hemianopsia, and pituitary stalk compression can cause panhypopituitarism (eg, growth failure, pubertal delay).

11981

- Posterior fossa tumors typically present with signs of cerebellar dysfunction (eg, ataxia) and increased intracranial pressure (eg, headache, vomiting, papilledema).
- In children, the most common posterior fossa tumors are pilocytic astrocytoma and medulloblastoma.

3665

- Parinaud syndrome occurs in most patients with pineal gland masses (pinealomas) and presents as limitation of upward gaze, bilateral eyelid retraction, and light-near dissociation.
- Pineal gland tumors can also cause persistent headache and vomiting due to obstructive hydrocephalus.

18410

- Headaches, increased intracranial pressure, and an unprovoked first seizure are concerning for a brain tumor.
- Frontal lobe tumors may cause personality changes, abulia, and anhedonia.

2620

- Lung cancer is the most common neoplasm to metastasize to the brain and may present with headache, focal neurologic dysfunction, cognitive change, or seizure in the absence of pulmonary symptoms.
- MRI of the brain typically reveals multiple, well-circumscribed lesions with vasogenic edema at the gray-white matter junction.

22118

- Exposure to ionizing radiation is the greatest acquired risk factor for development of meningiomas, which can occur >20 years after exposure.

22120

- Meningiomas often occur at dural reflections and can cause headaches and progressive neurologic defects that localize to the brain area compressed by the mass.

106023

- A lesion impacting the medulla can lead to motor weakness with upper motor neuron signs and lower cranial nerve dysfunction (eg, hypoglossal nerve [CN XII]).
- If it is an obstructing mass, it can also cause symptoms of increased intracranial pressure.

4072

- Surgical resection is recommended for solitary brain metastasis in patients with good performance status and stable extracranial disease.

11997

- The presence of an extra-axial well-circumscribed dural-based mass that is partially calcified on neuroimaging is strongly suggestive of a meningioma.
- Meningiomas are considered benign primary brain tumors; however, they can present with headache, seizure, and focal neurologic deficits due to mass effect.
- In such cases, complete surgical resection is recommended.

22119

- Meningiomas are benign, very slow-growing tumors that may be asymptomatic.
- In asymptomatic cases, especially in elderly patients or those with comorbidities, observation and serial imaging are appropriate.

4311

- Patients with an intracranial mass often have headache associated with nausea and vomiting and/or focal neurologic deficits. Symptoms are often worse at night and with body positioning that increases intracranial pressure. An MRI of the brain is usually diagnostic.

Pituitary tumors

4255

- Craniopharyngiomas are benign suprasellar tumors that present with visual defects, headache, and symptoms of pituitary hormonal deficiencies.

Pituitary apoplexy

17504

- Thunderclap headache is a rapid-onset, severe headache often associated with nausea and abnormal mental status. When associated with hypotension (central adrenal insufficiency), bilateral visual field defects (compression of the optic chiasm), and ophthalmoplegia (oculomotor nerve [CN III] palsy), it suggests pituitary apoplexy.